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Editor-in-Chief, Journal of Bangladesh College of Physicians and Surgeons, BCPS Bhaban, 67, Shaheed Tajuddin Ahmed Sarani Mohakhali, Dhaka-1212, Tel : 8825005-6, 8856616, 9884189, 9884194, 9891865 Ext. 124, Fax : 880-2-8828928, E-mail : <bcps@bcps-bd.org> Editor's e-mail: journal.bcps@gmail.com

Journal of Bangladesh College of Physicians and Surgeons (JBCPS)

INFORMATION FOR AUTHORS

MANUSCRIPT PREPARATION AND SUBMISSION

Guide to Authors

The Journal of Bangladesh College of Physician and Surgeons **,** provides rapid publication (three monthly) of articles in all areas of the subject. The Journal welcomes the submission of manuscripts that meet the general criteria of significance and scientific excellence.

Papers must be submitted with the understanding that they have not been published elsewhere (except in the form of an abstract or as part of a published lecture, review, or thesis) and are not currently under consideration by another journal published by **INTERNATIONAL RESEARCH JOURNALS** or any other publisher.

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BCPS Bhaban
67 Shaheed Tajuddin Sarani
Mohakhali, Dhaka 1212,
BANGLADESH
Phone: +8802-8825005 +8802-8825005
Fax: +88002-8828928

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A manuscript number will be mailed to the corresponding author within two working days.

The **cover letter** should include the corresponding author's full address and telephone/fax numbers and should be in an e-mail message sent to the editor, with the file, whose name should begin with the first author's surname, as an attachment.

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Article Types

FIve types of manuscripts may be submitted:

Editorials: It will be preferably written invited only and usually covers a single topic of contemporary interest.

Original Articles: These should describe new and carefully confirmed findings, and experimental procedures should be given in sufficient detail for others to verify the work. The length of a full paper should be the minimum required to describe and interpret the work clearly.

Short Communications: A Short Communication is suitable for recording the results of complete small investigations or giving details of new models or hypotheses, innovative methods, techniques , images in clinical practice, letter to editors, short reports or apparatus. The style of main sections need not conform to that of original article . Short communications are 2 to 4 printed pages (about 6 to 12 manuscript pages) in length.

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Case Reports: This should cover uncommon and/or interesting cases with appropriate confirmation process.

Review Process:

All manuscripts are initially screened by editor and sent to selective reviewer. Decisions will be made as

rapidly as possible, and the journal strives to return reviewers' comments to authors within 3 weeks. The editorial board will re-review manuscripts that are accepted pending revision. The JBCPS editorial board will try to publish the manuscript as early as possible fulfilling all the rigorous standard journal needs.

I. A. Preparing a Manuscript for Submission to JBCPS
Editors and reviewers spend many hours reading manuscripts, and therefore appreciate receiving manuscripts that are easy to read and edit. Much of the information in this journal's Instructions to Authors is designed to accomplish that goal in ways that meet each journal's particular editorial needs. The following information provides guidance in preparing manuscripts for this journal.

Conditions for submission of manuscript:

- All manuscripts are subject to peer-review.
- Manuscripts are received with the explicit understanding that they are not under simultaneous consideration by any other publication.
- Submission of a manuscript for publication implies the transfer of the copyright from the author to the publisher upon acceptance. Accepted manuscripts become the permanent property of the Journal of Bangladesh College of Physicians and Surgeons and may not be reproduced by any means in whole or in part without the written consent of the publisher.
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Ethical aspects:

- Ethical aspect of the study will be very carefully considered at the time of assessment of the manuscript.
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- Permission of the patients and/or their families to reproduce photographs of the patients where identity is not disguised should be sent with the manuscript. Otherwise the identity will be blackened out.

Preparation of manuscript:
Criteria: Information provided in the manuscript are important and likely to be of interest to an international readership.

Preparation:

1. Manuscript should be written in English and typed on one side of A4 (290 x 210cm) size white paper.
2. Margin should be 5 cm for the header and 2.5 cm for the remainder.
3. Style should be that of modified Vancouver.
4. Each of the following section should begin on separate page :
   - o Title page
   - o Summary/abstract
   - o Text
   - o Acknowledgement
   - o References
   - o Tables and legends.

Pages should be numbered consecutively at the upper right hand corner of each page beginning with the title page

I. A. 1. a. General Principles

- The text of observational and experimental articles is usually (but not necessarily) divided into the following sections: Introduction, Methods, Results, and Discussion. This so-called "IMRAD" structure is a direct reflection of the process of scientific discovery.
- Long articles may need subheadings within some sections (especially Results and Discussion) to clarify their content. Other types of articles, such as case reports, reviews, and editorials, probably need to be formatted differently.
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- Authors need to work closely with editors in developing or using such new publication formats and should submit supplementary electronic material for peer review.
- Double-spacing all portions of the manuscript—including the title page, abstract, text, acknowledgments, references, individual tables, and

legends—and generous margins make it possible for editors and reviewers to edit the text line by line and add comments and queries directly on the paper copy.

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- Authors should number on right upper all of the pages of the manuscript consecutively, beginning with the title page, to facilitate the editorial process.

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Research reports frequently omit important information. Reporting guidelines have been developed for a number of study designs that JBCPS journals ask authors to follow. Authors should consult the Information for Authors of this journal. The general requirements listed in the next section relate to reporting essential elements for all study designs. Authors are encouraged also to consult reporting guidelines relevant to their specific research design. A good source of reporting guidelines is the EQUATOR Network (http: //www.equator-network.org/home/) or CONSORT network (http:// www.consort-statement.org).

I. A .2. Title Page

The title page should have the following information:

1. Article title. Concise titles are easier to read than long, convoluted ones. Titles that are too short may, however, lack important information, such as study design (which is particularly important in identifying type of trials). Authors should include all information in the title that will make electronic retrieval of the article both sensitive and specific.
2. Authors' names and institutional affiliations.
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9. The number of figures and tables. It is difficult for editorial staff and reviewers to determine whether he figures and tables that should have accompanied a manuscript were actually included unless the numbers of figures and tables are noted on the title page.

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To prevent potential conflicts of interest from being overlooked or misplaced, this information needs to be part of the manuscript. The ICMJE has developed a uniform disclosure form for use by ICMJE member journals (http://www.icmje.org/coi_disclosure.pdf) and JBCPS has accepted that.

I. A. 4. Abstract

- Structured abstracts are essential for original research and systematic reviews. structured abstract means introduction, methods, results and conclusion in abstract
- Should be limited to 250 words
- The abstract should provide the introduction of the study and blinded state and should state the study's purpose, basic procedures (selection of study subjects or laboratory animals, observational and analytical methods), main findings (giving specific effect sizes and their statistical significance, if possible), principal conclusions. It should emphasize new and important aspects of the study or observations. Articles on clinical trials should contain abstracts that include the items that the CONSORT group has identified as essential (http:// www.consort-statement.org).
- Because abstracts are the only substantive portion of the article indexed in many electronic databases, and the only portion many readers read, authors need to be careful that they accurately reflect the content of the article

I. A. 5. Introduction

- Provide a context or background for the study (that is, the nature of the problem and its significance). It should be very specific, identify the specify knowledge in the aspect, reasoning and what the study aim to answer.
- State the specific purpose or research objective of, or hypothesis tested by, the study or observation; the research objective is often more sharply focused when stated as a question.
- Both the main and secondary objectives should be clear.
- Provide only directly pertinent primary references, and do not include data or conclusions from the work being reported.

I. A. 6. Methods

The Methods section should be written in such way that another researcher can replicate the study.

I. A. 6. a. Selection and Description of Participants

- Describe your selection of the observational or experimental participants (patients or laboratory animals, including controls) clearly, including eligibility and exclusion criteria and a description of the source population. Because the relevance of such variables as age and sex to the object of research is not always clear, authors should explain their use when they are included in a study report—for example, authors should explain why only participants of certain ages were included or why women were excluded. The guiding principle should be clarity about how and why a study was done in a particular way. When authors use such variables as race or ethnicity, they should define how they measured these variables and justify their relevance.

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- Identify the methods, apparatus (give the manufacturer's name and address in parentheses), and procedures insufficient detail to allow others to reproduce the results. Give references to established methods, including statistical methods (see below); provide references and brief descriptions for methods that have been published but are not well-known; describe new or substantially modified methods, give the reasons for using them, and evaluate their limitations. Identify precisely all drugs and chemicals used, including generic name(s), dose(s), and route(s) of administration.
- Authors submitting review article should include a section describing the methods used for locating, selecting, extracting, and synthesizing data. These methods should also be summarized in the abstract.

I. A. 6. c. Statistics

- Describe statistical methods with enough detail to enable a knowledgeable reader with access to the original data to verify the reported results. When possible, quantify findings and present them with appropriate indicators of measurement error or uncertainty (such as confidence intervals).
- Avoid relying solely on statistical hypothesis testing, such as *P* values, which fail to convey important information about effect size. References for the design of the study and statistical methods should be to standard works when possible (with pages stated).
- Define statistical terms, abbreviations, and most symbols.
- Specify the computer software used.

I. A. 7. Results

- Present results in logical sequence in the text, tables, and illustrations, giving the main or most important findings first. Please keep the result the sequence of specific objective selected earlier.
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- When data are summarized in the Results section, give numeric results not only as derivatives (for example, percentages) but also as the absolute numbers from which the derivatives were calculated, and specify the statistical methods used to analyze them.
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- Avoid nontechnical uses of technical terms in statistics, such as “random” (which implies a randomizing device), “normal,” “significant,” “correlations,” and “sample.” Where scientifically appropriate, analyses of the data by such variables as age and sex should be included.

I. A. 8. Discussion

- Emphasize the new and important aspects of the study and the conclusions that follow from them in the context of the totality of the best available evidence.
- Do not repeat in detail data or other information given in the Introduction or the Results section.
- For experimental studies, it is useful to begin the discussion by briefly summarizing the main findings, then explore possible mechanisms or explanations for these findings, compare and contrast the results with other relevant studies, state the limitations of the study, and explore the implications of the findings for future research and for clinical practice.
- Link the conclusions with the goals of the study but avoid unqualified statements and conclusions not adequately supported by the data. In particular, avoid making statements on economic benefits and costs unless the manuscript includes the appropriate economic data and analyses. Avoid claiming priority or alluding to work that has not been completed. State new hypotheses when warranted, but label them clearly as such.

I. A. 9. References

I. A. 9. a. General Considerations Related to References

- Although references to review articles can be an efficient way to guide readers to a body of literature, review articles do not always reflect original work accurately. Readers should therefore be provided with direct references to original research sources whenever possible.
- On the other hand, extensive lists of references to original work of a topic can use excessive space on the printed page. Small numbers of references to key original papers often serve as well as more exhaustive lists, particularly since references can now be added to the electronic version of published papers, and since electronic literature searching allows readers to retrieve published literature efficiently.
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- Authors are responsible for checking that none of the references cite retracted articles except in the context of referring to the retraction. For articles published in journals indexed in MEDLINE, the ICMJE considers PubMed the authoritative source for information about retractions.

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- References should be numbered consecutively in the order in which they are first mentioned in the text.
- Identify references in text, tables, and legends by Arabic numerals in superscript.
- References cited only in tables or figure legends should be numbered in accordance with the sequence established by the first identification in the text of the particular table or figure.

I. A. 10. Tables

- Tables capture information concisely and display it efficiently.

- Use tables /fig that are relevant to study
- Try to limit the number of tables/figure
- Type or print each table with double-spacing on a separate sheet of paper. Number tables consecutively in the order of their first citation in the text and supply a brief title for each.
- Do not use internal horizontal or vertical lines. Give each column a short or an abbreviated heading. Authors should place explanatory matter in footnotes, not in the heading. Explain all nonstandard abbreviations in footnotes, and use the following symbols, in sequence:

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- Identify statistical measures of variations, such as standard deviation and standard error of the mean.
- Be sure that each table is cited in the text. If you use data from another published or unpublished source, obtain permission and acknowledge that source fully.

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- Figures should be either professionally drawn and photographed, or submitted as photographic-quality digital prints. In addition to requiring a version of the figures suitable for printing,

  (for example, JPEG / GIF)
- Authors should review the images of such files on a computer screen before submitting them to be sure they meet their own quality standards. For x-ray films, scans, and other diagnostic images, as well as pictures of pathology specimens or photomicrographs, send sharp, glossy, black-and-white or color photographic prints, usually 127 _ 173 mm (5 _ 7 inches)
- Letters, numbers, and symbols on figures should therefore be clear and consistent throughout, and large enough to remain legible when the figure is reduced for publication.
- Photographs of potentially identifiable people must be accompanied by written permission to use the photograph. Figures should be numbered consecutively according to the order in which they have been cited in the text.
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- Measurements of length, height, weight, and volume should be reported in metric units (meter, kilogram, or liter) or their decimal multiples.
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- Use only standard abbreviations; use of nonstandard abbreviations can be confusing to readers.
- Avoid abbreviations in the title of the manuscript.
- The spelled-out abbreviation followed by the abbreviation in parenthesis should be used on first mention unless the abbreviation is a standard unit of measurement.

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- If a paper version of the manuscript is submitted, send the required number of copies of the manuscript and figures; they are all needed for peer review and editing, and the editorial office staff cannot be expected to make the required copies.
- Manuscripts must be accompanied by a cover letter, conflicts of interest form, authorship and declaration, proforma of which is available in JBCPS web site.

Editing and peer review: All submitted manuscripts are subject to scrutiny by the Editor in-chief or any member of the Editorial Board. Manuscripts containing materials without sufficient scientific value and of a priority issue, or not fulfilling the requirement for publication may be rejected or it may be sent back to the author(s) for resubmission with necessary modifications to suit one of the submission categories. Manuscripts fulfilling the requirements and found suitable for consideration are sent for peer review. Submissions, found suitable for publication by the reviewer, may need revision/ modifications before being finally accepted. Editorial Board finally decides upon the publishability of the reviewed and revised/modified submission. Proof of accepted manuscript may be sent to the authors, and should be corrected and returned to the editorial office within one week. No addition to the manuscript at this stage will be accepted. All accepted manuscripts are edited according to the Journal's style.

Submission Preparation Checklist

As part of the submission process, authors are required to check off their submission's compliance with all of the following items, and submissions may be returned to authors that do not adhere to these guidelines.

Check Lists

Final checklists before you submit your revised article for the possible publication in the Journal of Bangladesh College of Physicians and Surgeons:

1. Forwarding/Cover letter and declaration form
2. Authorship and conflicts of interest form
3. Manuscript
    - o Sample of the above documents is available in the following links: http://www.bcpsbd.org (registration required for download)
    - o If you have submitted mention document (1, 2, 3) above, when you first submitted your article then you don't need to re-submit but if there is change in the authorship or related then you have to re-submit it.

- General outline for article presentation and format
    - Δ Double spacing
    - Δ Font size should be 12 in arial
    - Δ Margins 5 cm from above and 2.5 cm from rest sides.
    - Δ Title page contains all the desired information (vide supra)
    - Δ Running title provided (not more than 40 characters)
    - Δ Headings in title case (not ALL CAPITALS, not underlined)
    - Δ References cited in superscript in the text without brackets after with/without comma (,) or full stop (.)
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    - Δ Numerals from 1 to 10 spelt out
    - Δ Numerals at the beginning of the sentence spelt out
- **Tables and figures**
    - Δ No repetition of data in tables/graphs and in text
    - Δ Actual numbers from which graphs drawn, provided
    - Δ Figures necessary and of good quality (colour)
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- **Title**
    - Δ Complete title of your article
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    - Δ Mention conflict of interest if any

- **Abstract**
  - Δ Do not use subheadings in the abstract
  - Δ Give full title of the manuscript in the Abstract page
  - Δ Not more than 200 words for case reports and 250 words for original articles
  - Δ Structured abstract (Including introduction, methods, results and discussion, conclusion) provided for an original article and (Introduction, results and discussion , conclusion) for case reports.
  - Δ Key words provided – arrange them in alphabetical order (three – five)
- **Introduction**
  - Δ Word limit 150 -200 words
  - Δ Pertinent information only
- **Material and Methods**
  - Δ Study Design
  - Δ Duration and place of study
  - Δ Ethical approval
  - Δ Patient consent
  - Δ Statistical analysis and software used.
- **Result**
  - Δ Clearly present the data
  - Δ Avoid data redundancy
  - Δ Use table information at the end of the sentence before full stop between the small bracket
- **Discussion**
  - Δ Avoid unnecessary explanation of someone else work unless it is very relevant to the study
  - Δ Provide and discuss with the literatures to support the study
  - Δ Mention about limitation of your study
- **Conclusion**
  - Δ Give your conclusion
  - Δ Any recommendation
- **Acknowledgement**
  - Δ Acknowledge any person or institute who have helped for the study
- **Reference**
  - Δ Abide by the Vancouver style
  - Δ Use reference at the end of the sentence after the full stop with superscript
- **Legends**
  - Δ Table
  - Δ Figures

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Vol. 32, No. 1, Page 1-58 **January 2014**

CONTENTS

EDITORIAL

Postgraduate Medical Education : An Overview

At Dhaka Post graduate Medical education was introduced in 1940's with establishment of School of tropical Medicine and Hygiene. This offered Diploma in Tropical Medicine of university of London. In 1961 Dhaka University Ordinance for Post Graduate courses were adopted. Courses in both Basic Medical science and Clinical Science were approved under Faculty of Medicine.[1] The College of Physicians and Surgeons Pakistan was established in **1962.** In 1965 Institute of Post Graduate Medicine and Research started functioning. In 1973 Faculty of Post Graduate Medical science and research started as a separate entity. National Professor Nurul Islam was the first Dean. Government of the Peoples' Republic of Bangladesh constituted an ad hoc committee of the College of Physicians and Surgeons of Bangladesh with Professor K S Haque as the President in 1972.[2] All fellows of the College of Physicians and Surgeons of Pakistan who hailed from Bangladesh were admitted as the founder fellows. With great endeavors and untiring efforts, these people were able to hold the first examination in July 1972. Bangabandhu Sheikh Mujib Medical University is the first and only medical university in Bangladesh, established in 1998.[3]

Though Royal College of Physicians of London was establishment in 1518, it took long time before the concept of postgraduate medical education started to bloom. Postgraduate medical education association was set up in UK in 1911 and Sir William Osler became its President. Osler in a valedictory address at McGill said "The hardest conviction to get into the mind of a beginner is that the education upon which he is engaged is not a college course, not a medical course but a life course, for which the work of a few years under teacher is but a preparation.[4]

Postgraduate medical education is based on adequate undergraduate training. Advanced learning necessarily demands sound understanding of the basics of the specialty. This again depends on the input in the medical colleges. Therefore postgraduate medical education cannot be considered in isolation. Considering the scenario of mushrooming of medical colleges without enough teachers, it is expected that outcome of medical graduates will fall short. But there will be students and institutes out performing and the post graduate program should have the ability pickup those students, otherwise long time & efforts in training of under qualified doctors may go in vain.

Once decided whom to train, next comes the question of how to do it. Colleges and educational bodies around the world decided on certain issues, like Professionalism, Humanism, skill, knowledge, and making learners ability of self learning and become a lifelong learner. Role of training institutes remains to provide requirements for expected outcome, providing opportunity to learn & assessing their performance time to time. At the same time degree/ diploma giving authority has to ensure the availability of resources and opportunities to the students of these training institutes by continuous monitoring.

The problems of medical education in a developing economy are many. Since the number of seats for the post graduation courses are very limited compared to the number of students being graduated, the students pay lesser attention on the practical training and more emphasis on the theoretical knowledge to score high in the PG entrance exams and part I exam as a result they are often found deficient in the performance of clinical skills and problem-solving which form the core of clinical competence. [8] Students fail to acquire the clinical skills, leadership qualities and human resource management to their maximum potential thus affecting the quality of doctors being produced. Assessment system, lack of facilities in the undergraduate program also contributes to this.

This is in sharp contrast to system followed in USA, where students need to pass a practical exam of United States Medical Licensing Examination (USMLE) step 2 Clinical Skills (CS) [5] Thus students need to have effective practical clinical skills before they can get license to practice in USA or to be eligible for admission to residency for higher education.

Specialist training mostly is in the traditional apprenticeship style rather than an appraisal based approach. Selection of assessment tools is not always governed by modern educational theory. Training in research, ethical issues, concepts of team work, and management is variable. Standards for accreditation are ill defined and not uniformly applied. Some institutes, however, have initiatives to meet these challenges.

The Accreditation Council for Graduate Medical Education (ACGME) in the USA has identified six learning outcomes for postgraduate medical education: patient care, medical knowledge, interpersonal and communication skills, professionalism, practice based learning and improvement, and system based practice.

World Federation for Medical Education (WFME)[6] recommends a set of global standards in postgraduate medical education structured according to 9 areas: 1. Mission and Outcomes, 2. Training Process, 3. Assessment of Trainees, 4. Trainees, 5. Staffing, 6. Training Settings and Educational Resources, 7. Evaluation of Training Process, 8. Governance and Administration and 9. Continuous Renewal. Standards are specified for each sub-area using two levels of attainment: Basic standard and Standard for quality development. **?**

The Gold Guide: A Guide to Postgraduate Specialty Training in the UK (the "Gold Guide") sets out the arrangements for the introduction of competence based specialty training in the UK . The development of this Guide has been through an iterative process of reflection and discussion using the Postgraduate Deans, Medical Royal Colleges and Faculties, professional associations and the health departments. The Postgraduate Medical Education and Training Board (PMETB) now incorporated within GMC since 2010 in UK, has very clearly described in its recommendations about the training of tomorrows doctor. [7]

While describing postgraduate medical education, R M Harden[8] mentioned four themes: the curriculum, the application of learning technologies, assessment of competence, and professionalism in medical education. However, to change our approach to teaching and learning is not easy for we must leave behind part of ourselves and our personal past experiences as trainees and trainers.

Patil[9]summarises the features of postgraduate training which includes : a progressive syllabus that has both formal and informal elements, a recognized trainer and training unit, proactive supervision, a balance of clinical duties and educational activities, protected time for education, and defined exit outcomes.

Outcome based education: A key trend in postgraduate medical education is a move to a model in which the emphasis has changed to focus on the product and the expected learning outcomes.[10] In outcome based education, the learning outcomes are clearly specified and decisions about the content of training and how it is organized, the educational strategies to be adopted, the teaching methods, the assessment procedures, and the educational environment are made in the context of the stated learning outcomes.[11]

A unitary approach to medical education: In 1973 Medearis and Kinney[12] argued convincingly that "medical education must be decompartmentalised and redesigned as a true continuum extending from secondary school through college, medical school, hospital training and postgraduate medical education". The various stages in the educational program will be integrated with the student and trainee advancing from one stage to the next and the exit learning outcomes of one phase being the entry requirements for the next.

Learning technologies: New learning technologies is a major area where we will see significant changes in postgraduate medical education in future. By using simulators learning is facilitated through the provision of effective feedback ,repetitive practice, a range of difficulty, multiple learning strategies, clinical variation, a controlled learning environment, and individualized learning.

E learning: Harden [13]highlighted some of the myths associated with e learning and concluded that e learning is not just a passing fad, it is not only about knowledge transfer, on line learning can be effective and efficient, students need not learn in isolation but can be part of an on line community, teachers and trainers have important but different roles, and technology may be queen but pedagogy is king. Khan Academy in USA is an example.

Assessment & feedback : We will see in the future more performance based assessment including more innovative and greater use of the OSCE. Training should

include workplace-based assessment. Recommendation of GMC of UK is as follows: (a) systematic observation of clinical practice, (b) direct observational procedure, (c) video, (d) judgments of multiple assessors, (e) consulting with simulated patients, (f) case record review, including OPD letters, (g) case-based discussion, s(h) oral presentations, (i) 360º peer assessment , (j) patient feedback surveys, (k) audit projects, (l) critical incident review. Outcomes from assessments must be used to provide feedback to the trainees on the effectiveness of the education and training.[14]

Faculty development: Newer ideas and methods of teaching and learning skill need to be incorporated in the training programs. Though in the advanced countries Department of Medical education is an integral part, it is yet to be in practice in our country. Center for Medical education has been playing some role but that is not adequate. In India National teacher training centers (NTTC) were established in 1974 at many medical colleges & postgraduate institutes. [15]NTTC activities includes six- to ten-day programs for medical educators with topics on education objectives, curriculum design, teaching methods, and assessment. Faculty-training courses facilitated the introduction of some innovations in various medical colleges in India and fostered the development of medical education units in other colleges. The USA based Foundation for Advancement of International Medical Education and Research (FAIMER)[16] is also supporting faculty development in India. Few of our faculties completed their online diploma. All doctors with particular responsibility for training will need to demonstrate evidence that they have acquired the necessary additional skills.

Conclusions:

Postgraduate Medical education in Bangladesh like many developing countries in Asia & Africa has its own problems. Lack of coordination among various stakeholders, inadequate number of trainers, poor faculty development program, deficiencies in the undergraduate program, lack of effective curriculum to focus on the structure and outcome of training are few of many.

What is the best way to solve the problems and ensure higher standard of tomorrows specialists? The solution probably lies in following the path of those who have solved their problem. A National commission for postgraduate medical education need to be established with representation from, Medical University, BCPS, Medical & Dental Council, Ministry of Health and BMA. This commission will review present problems, program of advanced countries and of the developing countries in the region and shall produce a National Guideline. There is no shortcut of structured training program. A National Medical Teachers Training Institute should be established and criterion to become a medical teacher should include training in medical education even if a short one. Like the students, teachers also need to become lifelong learners and that need to be ensured. We are now at the cross road of seeing a growing gap between what is possible educationally and what is delivered. It is clear that we need a new paradigm for postgraduate medical education. Outcome based program with adequate training and faculty development probably does not have any alternative. In delivering this, all stakeholders have a contribution to make and must accept a measure of responsibility for what happens in the future.

It is important that loyalty to the past does not cloud our minds or prevent us from sharing in the joy and satisfaction with what will be possible in the years ahead, if we allow it to happen. The comfort of inertia is not an option, for we need to face up to the annoyance or excitement.[8]

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Prof. Humayun Kabir Chowdhury
Prof. of Surgery
BIRDEM Hospital, Dhaka

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Clinicopathological Profile of Wilms' Tumour in Children

M MAZUMDER[a], A ISLAM[b], N FAROOQ[c], M ZAMAN[d]

Summary:

Introduction: Wilms' tumor is the most common primary malignant renal tumor of childhood. It is important to pick up the children with wilms' tumor earlier as early stages has excellent outcomes after treatment.

Objective : To find out the common clinical presentations and pathological profile of Wilms' tumor in children.

Methods and Materials : A hospital based prospective study done with twenty diagnosed patients of Wilms tumour enrolled from department of Pediatric haemato-oncology, BSMMU, Dhaka in the period between January to December 2008.

Results- The peak incidence of Wilms' tumor was in 1 to 5 years age group (80%,n=16). Median age at presentation was 49 months with male: female ratio 1.8:1.The most common presentation was abdominal swelling (80%,n=16),followed by flank mass (75%,n=15), abdominal pain (55%,n=11), haematuria (15%,n=3), hypertension (10%,n=2). Thirteen raised from right kidney, ratio of right to left involvement 1.8:1. Histologically 13(65%) patients had triphasic histology having blastemal, stromal and epithelial elements, 7(35%) was biphasic having blastema and epithelia. All had favourable histological pattern. Most patients presented in stage III (55%,n=11) followed by stage II (25%,n=5), Stage IV(10%,n=2), Stage I(10%,n=2). No bilateral presentation.

Conclusions : Most of the patients of Wilms' tumor presented within 1 to 5 years of age(80%) with abdominal distension(80%) and flank mass(75%), few associated with haematuria(15%) and hypertension(10%). Histologically all were favourable and maximum presented in stage III(55%) followed by stage II(25%).

Keywords: Clinicopathological profile, Histological pattern, Wilms' tumour.

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Introduction:

In 1899 surgeon Max Wilms (1867-1918) described seven children suffering from nephroblastoma in a monograph of 'mixed tumors'. It is now recognized that Wilms' tumor accounts for approximately 6% of pediatric cancers and is the second most common malignant abdominal tumor in childhood.[1] With multimodality treatment and recent advancement it has become one of the curable tumor.[2] With the introduction of adjuvant chemotherapy and sometimes radiotherapy, survival rates approaching 90% were achieved for localized tumor by 1970s.[3]

Wilms' tumor is a mixed embryonal neoplasm composed of three elements – blastema, epithelia and stroma. It may arise in one or both kidneys; the incidence of bilateral Wilms' tumor is 6%. It may be associated with hemihypertrophy, aniridia and genitourinary anomalies, including hypospadias and cryptorchidism, some syndromes eg. WAGR or Denys-Drash syndromes.[1] The incidence of wilms' tumor is approximately 8 cases per million children <15 yrs of age.[1] It usually occurs in children between 2-5 yrs of age. The tumor presents at an earlier age among boys , with the mean age at diagnosis for those with unilateral disease being 41.5 months compared with 46.9 months among girls.[4] The median age is highest for patients with unilateral unicentric disease (36.1 mon) and lowest for those with synchronous bilateral Wilms' tumors (25.5 mon).[5] Wilms' tumor usually presents as an abdominal mass often noticed by the parents during bathing or dressing. The mass is usually smooth, firm, of variable size and

a. Dr. Monika Mazumder, Registrar (Neonatology), Department of Paediatrics, Rangpur Medical College Hospital, Rangpur.

b. Prof. Afiqul Islam, Chairman, Department of Pediatric Haemato-oncology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka.

c. Dr.Nusrat Farooq, Registrar (Neonatology), Apollo Hospital, Dhaka.

d. Dr.Mahmuda Zaman, Registrar (paediatrics), Apollo Hospital, Dhaka.

Address of Correspondence: Dr. Monika Mazumder, Registrar (Neonatology) Department of Paediatrics, Rangpur Medical College Hospital Rangpur, Mobile: 01920703009, 01711134991, E-mail: monikamz26@yahoo.com / monika.bd@in.com,

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occasionally may cross midline. Abdominal pain in about 30%- 40% cases, vomiting, constipation, haematuria(12–25%), hypertension (3- 25%), pallor, signs of Wilms' tumor associated syndromes eg. aniridia, facial dysmorphism, partial or complete hemi-hypertrophy, hypospedias, cryptorchidism, pseudo-hermaphroditism.[1, 6] Wilms'tumor (hereditary or sporadic) appears to result from changes in one or more of at least ten genes. The W*T1*, WT2, p53.[4,7]Favorable histology (90%) having blastema, stroma, epithelia without any anaplastic features has cure rate close to 90%. Unfavourable histology (10%) is characterized by the presence of anaplasia.[1]There is no statistical record on incidence of Wilms Tumour in Bangladesh, though a number of cases are found in different hospitals each year. In a survey of surgical problems in children in Dhaka Shishu Hospital from 1981 to 1990, it was found that out of 12189 patients, 228 patients (about 2%) were diagnosed as Wilms' Tumour.[8] Another study found that out of 90 cases of malignancy, 22% was nephroblastoma during the four year study period.[9]

Development of surgical techniques, sensitivity to radiation and availability of several active chemotherapeutic agents led to a dramatic change in prognosis for most patients with this once lethal malignancy. So the findings of this study could help the health professionals in early diagnosis of Wilms' tumor and offer the patients the best options of treatment that brings in much better outcome.

Methods and materials:

A hospital based prospective study done with twenty diagnosed patients of Wilms tumour enrolled from department of Pediatric Haemato-oncology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka in the period between January to December 2008.Age below 15 years and diagnosed case of Wilms' tumor as suggested by history, clinical examinations with or without imaging was included. Age over 15 years and renal mass due to other cause was excluded. Objective was to find out the common clinical presentations and pathological profile of Wilms' tumor in children. During the study period total 23 patients were enrolled. Among them, 2 cases were neuroblastoma and 1 case was renal cell carcinoma as diagnosed later by histopathology.

Results:

In this study 20 patients were enrolled and the findings are subsequently presented in tables and figures.

Table-I

Age distribution of study patients.

	Age in months			Age in years		Median age (months)
	0 - 12	13 - 24	25 – 60	5 - 10	11 - 15	49
No of patients	1	3	12	2	2	
percentage	5%	15%	60%	10%	10%	

Table 1 shows age distribution of study patients. Median age was 49 months. 16 (80%) presented within 5 yrs of age and 12 (60%) presented between 25–60 months of age.

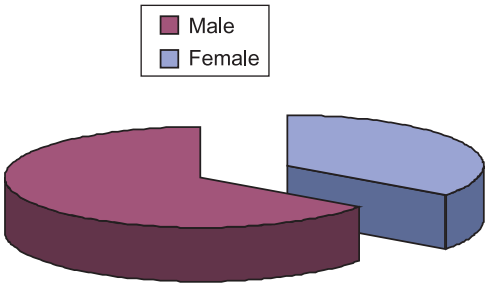

Fig.-1: *Sex distribution of study patients (n-20).*

Fig.-1: The pie chart shows sex distribution of study patients. 13(65%) patients were male, 7(35%) patient were female. Male : Female =1.8:1.

Table-II

Clinical presentation of study patients(n=20).

Clinical presentation	No. Of patients	Percentage (%)
Abdominal swelling	16	80%
Flank mass	15	75%
Abdominal pain	11	55%
constipation	8	40%
Vomiting	6	30%
Pallor	4	20%
Haematuria	3	15%
Hypertension	2	10%

Table 2 showing distribution of percentage of presenting features in study group.The most common presentation was abdominal swelling (80%, n=16) followed by flank mass(75%, n=15), abdominal pain(55%, n=11), haematuria(15%, n=3), hypertension(10%, n=2).

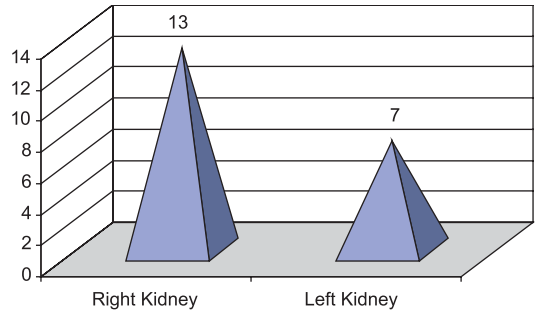

Fig.-2: *Site of origin of Wilms' tumor.*

Bar diagram showing origin of Wilms tumor by ultrasonography. Thirteen (65%) raised from right kidney, 7(35%) from left kidney.There was no bilateral tumor.R:L = 1.8 : 1.

Table-III

Biopsy and Histopathology findings (n= 20).

Histology	No. of patients	%
Blastemal cells (Small rounded cells scanty blue cytoplasm, hyperchromatic neuclei)	20	100%
Epithelial cells(forming tubules, cords, rosette)	16	80%
Stromal elements	13	65%
Necrosis	6	30%

Table-3 showing all patients had blastemal cells, 16(80%) patients had epithelial cells and 13(65%) patients had stromal elements in histopathology. Necrosis was found in 4(20%) patients. None had anaplasia.

Bar diagram showing 11(55%) was in stage III, 5(25%) in stage II, stage I and stage IV was 2(10%) each.

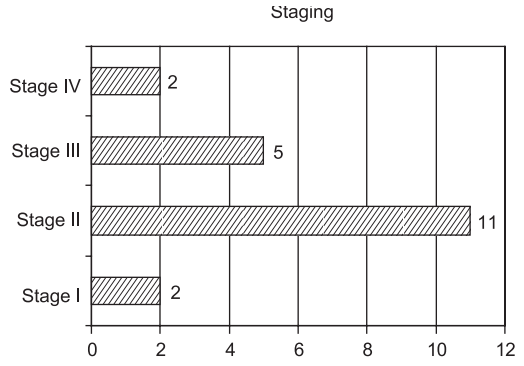

Fig.-3 : *Staging of Wilms tumor in study patients(n=20).*

Discussion:

Wilms' tumor is the commonest primary renal neoplasm in children. Most children have non specific symptoms in early disease course. So the tumor is often missed unless there is high suspicion and good abdominal examination in early stage. . It is important to pick up the children with wilms' tumor earlier as early stages have excellent outcomes after treatment.

This study is a simple prospective study reflecting the mode of clinical presentation and histopathological pattern in patients of Wilms' tumor in children under 15 years. The study was carried out in the department of Pediatric Haemato oncology, BSMMU, from January to December 2008.

Twenty three cases were enrolled primarily. History taking included history of consanguinity, family history, and onset of the disease, complaining symptoms, presence of congenital anomalies and the method of surgical interference like FNAC. Clinical examination included examination of any congenital anomalies as aniridia, genitourinary malformations, hemi-hypertrophy or signs of overgrowth and hypertension. Radiological examination included chest X-ray (CXR), abdomen-pelvic ultrasonography and CT scan for confirming the origin and any residual or metastatic disease. Laboratory examination included complete blood picture (CBC), urinalysis, renal and liver profiles, and diagnosis was confirmed by biopsy and histopathology. Among the 23 cases 2 cases were neuroblastoma and 1 case was renal cell carcinoma as diagnosed later by histopathology. So twenty patients were finally diagnosed as Wilms' tumor.

Among the twenty (20) cases of Wilms tumor 16 (80%) presented below 5 years of age and 12 (60%) was within 25 – 60 months with male: female= 1.8:1. Median age at presentation was 49 months. The results of the study almost corroborate with the study by Paul et al, who found 90% presented in<6 years and 66.7% in <3 years with a Male: Female= 2.1:1.[10] The study of Hisham et al, *2005*showed the male : female ratio 1.5 :1.[11]

Major clinical presentations were abdominal swelling (80%, n=16), flank mass (75%, n=15), abdominal pain (55%, n=11), constipation (40%, n=8), vomiting (30%, n=6). Few patients had haematuria (15%, n=3), hypertension (10%, n=2). Some patient also presented with gradual pallor. None of them had syndromes or any congenital anomaly. In the study of Hisham etal, the most common complaint was abdominal swelling (82.3%), followed by haematuria (14.5%), then abdominal pain (13%).[11] The work of Pianezzaet al,

where an abdominal mass was the most common presenting feature (85%), followed by abdominal pain (17%), and then haematuria (10%).[12] Both studies almost corroborate with the findings of present study in term of clinical presentation.

Of the 20 cases 65% (n=13) was originated from right kidney and 35%(n=7) from left kidney, Right: Left = 1.8:1. This corroborates with the study of K Basu et al, as they found right kidney involved in 63% cases and left kidney 37% cases.[13] There was no bilateral tumor. But in the study of Sharma et al they found left kidney involvement in 60% cases and Hung IJ et al found 55.8% right kidney and 38.2% left kidney, 6.8% bilateral involvement.[14] The work of Pianezza et al, also reported 4.8% cases as bilateral Wilms' tumor.[12]

Histological results showed triphasic(having blastema, epithelia, stroma) histology in 65%(n=13) cases, biphasic (blastema and epithelia) in 35%(n=7) cases. None had anaplastic or monophasic histology. So all cases (100%) were of favourable type according to prognostic consideration. This findings corroborates with the study of Basu et al.[13] But the study of Quijano and Drut found blastemal cells in all cases but stromal cells in 90% cases and epithelial cells in only 40% cases.[15] Hung IJ et al found 85.3% favourable histology and also Hisham et al reported 65.4% favourable histology.[11, 14] These two studies does not corroborate with present study.

According to National Wilms' Tumor Study Group (NWTSG) patients were divided into 5 stages.In this study most patients presented in stage III (55%,n=11), followed by stage II (25%,n=5), and 10%(n=2) each in stage I and IV. No one was in stage V. This is comparable with the study of K Basu et al where they found 68% cases in stage III fillowed by 21% stage II and 10.5% stage IV.[10] But Hung IJ et al found 43.2% in stage I, 23% siage III, 19.3% stage II and 6.8% each in stage IV and V. In the study of Hisham et al, 22 patients (35.5%) had stage I disease, 17 cases (27.4%) had stageII, 16 cases (25.8%) had Stage III, 4 cases (6.5%) had stage IV and 3 cases (4.8%) had stage V disease.[11]To describe clinicopathological profile of Wilms' tumor, a study period of one year may not reflect the real situation. As per consequence sample size was also small. An extended period follow up study may reveal the actual situation and clinicopathological correlation with outcome in patients with Wilms' tumor. So this study raises the need for more large scale work.

Conclusion:

Most of the patients of Wilms' tumor presented within 1 to 5 years of age(80%)with abdominal distension (80%) and flank mass(75%), few associated with haematuria(15%) and hypertension(10%). Histologically all were favourable and most presented in stage III(55%) followed by stage II (25%). So the patients of Wilms' tumor presents mostly in later stages. Data obtained from this study may lead to early detection due to early clinical suspicion and thus further improvement in prognosis of paediatric malignant solid tumor in our country.

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Cholestatic Jaundice in Infants – An Experience in Tertiary Care Hospital

FR CHOWDHURY[a], K CHOWDHURY[b], ASMB KARIM[c]

Summary:

Background: Neonatal cholestasis is defined as prolonged elevation of serum levels of conjugated bilirubin beyond the first 14 days of life. Cholestasis in a newborn can be due to infectious, genetic, metabolic, or undefined abnormalities giving rise to mechanical obstruction of bile flow or to functional impairment of hepatic excretory function and bile secretion. Early detection and timely accurate diagnosis are important for successful treatment and a favorable prognosis.

Objective: The present study has been designed to determine the etiology of cholestatic jaundice in infants along with their clinical profile.

Methodology: This cross-sectional study was conducted from August 2010 through January 2011 in the Paediatric Gastroenterology & Nutrition Department, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka. For the study purpose 40 consecutive cases of cholestatic jaundice were included who fulfilled the inclusion criteria.

Result: Biliary atresia was the commonest (42.5%) cause of cholestatic jaundice followed by neonatal hepatitis/ idiopathic neonatal hepatitis. Other causes of cholestatic jaundice were choledochal cyst and hypothyroidism. Most of the infants were term and of normal birth weight in cases of biliary atresia (BA) but in NH/INH group significant number of infants were preterm and of low birth weight. Mean age at onset of jaundice was 10.1 ± 4.18 days, and mean age at presentation was 113.7 ±15.38 days. In cases of BA Jaundice, intermittent / persistent pale stool, dark urine was found in all cases and hepatomegaly and splenomegaly were found in 88.2% and 64.8% of cases respectively. Ultrasonographically in most of the cases of BA gallbladder was found either small in size or absent or bile ducts were not visualized. In cases of NH/INH visualization of normal gallbladder while fasting and contraction was observed after meal. Histologically typical features BA were found in 12 out of 17 cases of BA and features of early biliary cirrhosis in 4 infants and 10 patients showed features of INH.

Conclusion: Biliary atresia was found to be the commonest cause of neonatal cholestasis in the present study.

Key Words: Neonatal cholestasis, Biliary atresia, Neonatal jaundice.

(J Banagladesh Coll Phys Surg 2014; 32: 9-15)

Introduction:

Neonatal cholestasis is defined as prolonged elevation of serum levels of conjugated

bilirubin beyond the first 14 days of life[1]. Conjugated hyperbilirubinemia is defined by a serum conjugated bilirubin concentration of greater than 1mg/dl (17.1mol/ l) if the total bilirubin is 5mg/dl (85.5mmol/l) or more than 20%. It is an abnormal finding and requires additional evaluation if it persist beyond 2 weeks of life[2].

a. Dr. Farzana Rahman Chowdhury, Registrar, Shahabuddin Medical College, Gulshan, Dhaka.

b. Dr. Kona Chowdhury, Assistant Professor, Department of Paediatrics, Ad-din Women's Medical College and Hospital.

c. Prof. ASM Bazlul Karim, Professor of Paediatric Nutrition and Gastgroenterology, Bangabandhu Sheikh Mujib Medical University, Dhaka.

Address of Correspondence: Dr. Farzana Rahman Chowdhury, Registrar, Shahabuddin Medical College, Gulshan, Dhaka. Flat # A2, DOMINNO Altura, 342, Segunbagicha, Dhaka-1000, Tel: 01715786329, E-mail: shomi9@yahoo.com

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An infant with cholestatic jaundice usually presents with prolonged jaundice, pale stool and dark urine. Acholic stool, a cardinal feature of cholestasis should be promptly evaluated.

Other clinical features depend on etiology of cholestasis. About 70% cases of neonatal cholestasis are due to biliary atresia & Idiopathic neonatal hepatitis (INH)[3]. Idiopathic neonatal hepatitis occurs more commonly in males, especially those born prematurely or with low birth weight; there is a familial incidence of approximately 10% to 20%[4]. In contrast, biliary atresia is more common in females of term and of normal birth weight and familial cases are rare. Affected children present with firm hepatomegaly and splenomegaly. Congenital malformations, including cardiac anomalies, polysplenia, intestinal malrotation and situs inversus, may be found in almost a third of infants with biliary atresia[5].

Initially, the symptoms of BA are indistinguishable from neonatal jaundice, due to other causes. Symptoms are usually evident between one and six weeks after birth[6].

Prolonged jaundice that is resistant to phototherapy and/or exchange transfusion should prompt a search for secondary causes. By this time, liver enzymes are generally measured, and these tend to be grossly deranged. Ultrasound investigation or other forms of imaging can confirm the diagnosis. Further testing includes radioactive scans of the liver and a liver biopsy[7].

The early detection of biliary atresia is one of the major challenges facing pediatrician when evaluating the jaundice in infant. Early recognition of liver disease greatly facilitates the care and outcome of infants. A key component of the work-up is measurement of serum conjugated bilirubin levels after 2 weeks which if elevated should prompt the clinician to initiate a work–up to determine the cause of neonatal cholestasis[8]. In general, if patient is developing progressive jaundice soon after birth and is still jaundiced at 2 weeks of life, or develops jaundice within 3 months of life, a work up for neonatal cholestasis should begin[9]. The success rate for establishing good bile flow after the Kasai operation is much higher (90%) if performed before 8 wks of life[10].

Nowadays, development of sophisticated diagnostic modalities and methods makes the diagnosis possible in early stages and the underlying cause could be easily discerned. In spite of this, unfortunately there are limited data about the disease among Bangladeshi infants.

As the outcome of biliary atresia depends on early recognition and timely surgery, so the study was undertaken in infants having jaundice developed after 2 weeks of life to determine the age of onset and to document the common clinical presentation.

Methodology

Study site & duration:

This hospital based cross sectional descriptive study was conducted at the Paediatric Gastroenterology Department, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka from August 2010 to December 2011.

Study population:

Admitted patient of the department were the study population and those who gave consent were enrolled.

Infants below 12 months of age are included with all of the following criteria:

1. Jaundice developed before 3 months of age and persisted for at least 2 weeks.
2. Intermitted or persistent pale colored stool.
3. Passage of dark urine
4. Conjugated bilirubin concentration more than 20% of total bilirubin or >2mg/dl

The following babies are excluded from this study:

- Very sick infants with features of liver failure.
- Parents not willing to participate in the study.
- Patient of hepatocellular carcinoma & hemolytic jaundice.

After enrolment patients underwent the following routine investigations:

Serum bilirubin, fractionated serum bilirubin, liver function tests like alanine aminotransferase (ALT), alkaline phosphatase, gamma glutamyl transferase and prothrombin time. Complete blood count, Blood glucose, reducing substance in urine, urine for routine microscopic examination and culture sensitivity, bacterial culture of blood, ultrasonography of liver and biliary system.

Investigations to establish a specific diagnosis: Endocrine studies (FT4, TSH), TORCH screening, VDRL, HBsAg, serum alpha 1 antitrypsin level, hepatobiliary scintigraphy (HIDA scan), percutaneous liver biopsy were done.

Data collection method: Data were collected using a preformed data collection sheet (questionnaire). Statistical analysis was done using the statistical package for social sciences (SPSS version 17.0 for Windows, SPSS Inc. Chicago, IL). All the values were expressed as Mean ± SD; Students unpaired t test were applied as statistical tools. p value of < 0.05 was considered as significance.

Results:

In this study a total of 40 children were studied. Biliary atresia was found to be the commonest, 17(42.5%), cause of neonatal cholestasis followed by idiopathic neonatal hepatitis, 10 (25%) and neonatal hepatitis, 08 (20%). Among the neonatal hepatitis, cyto-megalovirus, rubella virus and herpes simplex virus were found in 4, 2 and 2 cases respectively as identifiable causes. (Table I)

In this study the mean age at admission of biliary atresia was 113.7±15.38 days and that of neonatal hepatitis 105.05±16.81 days. The overall mean age at admission of cholestatic cases was 111.9±21.14 days. (Table II)

In this study twenty five children were male and fifteen female. Biliary atresia was present in 70.6% male & in 29.4% female childrens and INH was present in 55.5% male & 44.4% female children. (Table III)

The mean age at onset of jaundice was 10.1 ± 4.18 days and mean age at admission was 113.7 ± 15.38 days in case of biliary atresia. The mean age at onset of jaundice was 12.4 ± 4.7 days and mean age at admission was 105.05 ± 16.81 days in case of NH or INH. Thus an overall delay in seeking treatment was 103.6 days in biliary atresia and 92.65 days in NH & INH cases. (Table IV)

Among the studied patient 14 (82.3%) patients were term and 03 (17%) were preterm in biliary atresia case but 13 (72.2%) were term and 05 (27.8%) preterm in NH/ INH. Most of the patients in biliary atresia cases were term infants. In this study jaundice and dark urine were found in all cases of both biliary atresia and neonatal hepatitis or idiopathic neonatal hepatitis cases. Persistent acholic stool was an important finding of biliary atresia 15 (88.2%) cases and intermittent acholic stool was a significant finding of NH /INH, 14 (77.7%) cases. In biliary atresia 15 (88.2%) patients were found to have hepatomegaly and the liver was firm to hard in consistency. Fourteen (77.7%) patients with NH / INH were found to have hepatomegaly. Splenomegaly was commoner in NH /INH case than in biliary atresia. Ascites was seen in one patient with biliary atresia and in three cases of NH / INH. None of the patients showed eye findings like cataract, posterior embryotoxon, cherry red spot or chorioretinitis. (Table V)

In biliary atresia the mean serum total bilirubin was 12.4±2.55 mg/dl and that in NH/INH was 14.7±4.64 mg/dl and serum ALT wass 219.7±102.5 U/L and 423±92.6 U/L in biliary atresia and NH/INH cases respectively which was not statistically significant. The mean serum alkaline phosphatase was 1048.8±162.8U/L in biliary atresia and 629.6±160.8 in NH/INH cases. Gammaglutamyl transpeptidase, prothrombin time or serum albumin level showed no significant difference between biliary atresia and NH/INH groups. (Table VI) USG was done in all infants with cholestatic jaundice. In biliary atresia gall bladder was found either small in size or absent or bile channels were not visualized and no contraction of gall bladder was seen even after meals. On the contrary, in neonatal hepatitis, gall bladder was normally visualized with biliary channels and contraction of gallbladder was seen after meals. (Table VII).

HIDA scan was done in selected patients i.e. in fourteen infants. Nine patients showed delayed uptake but normal excretion, which is consistent with NH, Where as five infants showed normal uptake of the isotope but absent excretion into the biliary channels and intestine, which is consistent with biliary atresia.

Liver biopsy was done in twenty six infants. Typical features of biliary atresia were found in twelve patients. Ten patients showed features of idiopathic neonatal hepatitis and four patients had features of early biliary cirrhosis. Liver biopsy could not be done in other cases due to prolonged prothrombin time, huge ascites or lack of parental consent. (Table VIII)

Table-I

Etiology of cholestasis in studied patients (n=40)

Etiology	Number	Percentage
Biliary atresia	17	42.5
Neonatal hepatitis:	08	20
Cytomegalovirus(CMV)	04	
Rubella Virus	02	
Herpes simplex virus	02	
Idiopathic neonatal hepatitis	10	25
Miscellaneous:		12.5
Choledochal cyst	3	
Hypothyroidism with CMV	2	
Total	40	100

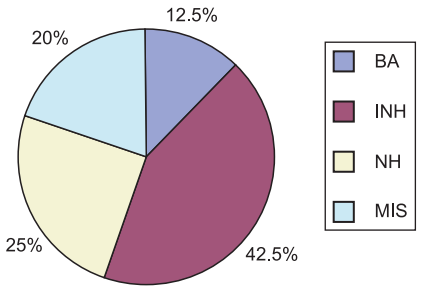

Fig.-1: *Etiology of cholestasis in studied patients (n=40)*

Table-II

Age distribution of studied patients at admission (n=40)

Diagnosis	No	Mean age (days)	Standard deviation	p-value
Biliary atresia	17	113.7	±15.38	0.12
NH & INH	18	105.05	±16.81	0.12
Miscellaneous	05	106.6	±20.4	
Total patients	40	111.9	±21.14	

Table-III

Sex distribution of studied patients (n=40)

Sex	Biliary Artesia	INH and NH	Miscellaneous	Total
Male	12 (70.6%)	10 (55.5%)	03 (60%)	25 (100%)
Female	05 (29.4%)	08 (44.4%)	02 (40%)	15 (100%)
Total	17	18	05	40

Table-IV

Age at onset of symptoms and age at admission.

	Biliary atresia (n=17) Mean ± SD	NH & INH (n= 18) Mean ± SD	p value
Age at onset (days)	10.1 ± 4.18	12.4 ± 4.7	0.73
Age at admission (days)	113.7 ± 15.38	105.05 ± 16.81	0.58
Delay (days)	103.6	92.65	

Table-V

Clinical features of studied cases.

Clinical characteristics	Biliary atresia(n=17) No. (%)	NH/ INH (n=18) No. (%)
Jaundice	17 (100)	18 (100)
Dark urine	17 (100)	18 (100)
Persistent acholic stool	15 (88.2)	04 (22.2)
Intermittent acholic stool	02 (11.8)	14 (77.7)
Hepatomegaly	15 ((88.2)	15 (83.3)
Splenomegaly	11 (64.7)	14 (77.7)
Ascites	01 (5.8)	03 (16.6)

NH = Neonatal hepatitis
INH = Idiopathic neonatal hepatitis

Table-VI

Biochemical parameters of studied cases.

Liver function tests	Biliary atresia(n=17) Mean ±SD	NH& INH(n=18) Mean+ SD	p-value
Serum total bilirubin (mg/dl)	12.4±2.55	14.7±4.64	0.04
Serum direct bilirubin (mg/dl)	7.2±1.6	11.0±4.0	<0.01
Serum albumin (gm/dl)	32.4±3.1	30.1±2.9	0.69
ALT(U/L)	219.7±102.5	423.9±92.6	0.99
Alkaline phosphatase (U/L)	1048.8±162.8	629.6±160.8	0.84
Gamma glutamyl transpeptidase (ãGT) (U/L)	624.9±129.5	553.0±111.2	0.52
Prothrombin time (sec) of patient	15±0.49	18.2±3.9	0.11
INR	1.44±2.7	1.5±0.43	0.28

Table-VII

Findings of Ultrasonography in studied patients (n=40)

Ultrasonography findings	Diagnosis	No
Small/absent gallbladder with non-visualized biliary channel	Biliary atresia	17
	INH / NH & Miscellaneous	20
Normal Gallbladder with visualized biliary channel	Cystic lesions were seen in biliary tree	03
Choledochal cyst		

Table-VIII

Liver biopsy findings in studied patients (n= 26)

Findings	Diagnosis	No.
Ductular proliferation, bile plugs, intraportal fibrosis	Biliary atresia	12
	Idiopathic neonatal hepatitis	10
Inflammation, hepatocyte necrosis and giant cell transformation	Biliary atresia with biliary cirrhosis	04
Ductular proliferation, bile plugs, inflammation, fibrosis		

Discussion:

This hospital based cross sectional study was carried out to determine the frequency of biliary atresia in infants admitted with cholestatic jaundice along with their clinical profile. During the study period a total of 40 infants were admitted in the Pediatric Gastroenterology and nutrition department of Bangabandhu Sheikh Mujib Medical University, Dhaka. In the present series of cholestatic jaundice, biliary atresia was found in 17 (42.5%) cases, neonatal hepatitis in 8 (20%) and idiopathic neonatal hepatitis in 10 (25%) cases.

A retrospective study was conducted among Bangladeshi infant to find out the etiology and clinical profile of neonatal cholestatic disorders[11]. A total of 62 infants with cholestatic jaundice were studied who developed jaundice before three months of age and persisted for more than two weeks. In that study biliary atresia was found in 16 (25.8%), neonatal hepatitis in 22 (35.5%) and idiopathic neonatal hepatitis in 15 (24.2%) cases. Neonatal hepatitis was the commonest cause of cholestatic jaundice in that study but biliary atresia was found to be the commonest cause in the present study. This difference may be due to small sample size.

The mean age at admission to hospital of biliary atresia cases was 113.7 ± 15.38 days, though the mean age of onset of jaundice was 10.1 ± 4.18 days and average delay was 103.6 days. Karim & Kamal[11] reported the mean age at presentation to hospital in their series was 105 days while the mean age at onset of jaundice was 5.8 days and the average delay was 99.2 days. These findings are almost consistent with findings of present study. Delay in diagnosis of cholestatic disorders especially biliary atresia is also a problem in developed countries[12]. If the treatment of extra hepatic biliary atresia is delayed beyond the first 60 days of life, the only option left thereafter is liver transplantation, which is not commonly feasible on a large scale in developing countries. This delay contributes to increase in morbidity and mortality and also to poor outcome[13].

Amongst the clinical feature only acholic stool was differentiating (88.2% BA vs. 22.2% NH). This was also represented by Karim & Kamal[11]. There was not much differentiating point as regard to other clinical features. Intermittent acholic stool found in biliary atresia may be due to the progressive obliterative cholangiopathy *i.e.* incomplete obliteration of entire extrahepatic biliary tree. Conversely in long standing cases of NH/INH persistent acholic stool usually found.

Common signs of the studied cases of biliary atresia were hepatomegaly (88.2%), splenomegaly (64.7%) and ascites (5.8%). In NH/INH cases hepatomegaly was found in 83.3% cases, splenomegaly in 77.7% cases and ascites in 6.6% cases.

Amongst the laboratory findings there were no such differentiating characteristics. However ALT and ALP was found to be raised in NH/ INH and BA respectively but they were not significant statistically.

In biliary atresia cases the mean serum total bilirubin was found to be 12.4±2.55 mg/dl and that in NH/INH cases 14.7±4.64 mg/dl, this value is statistically significant. Serum ALT was found 219.7±102.5 U/L and 423.9±92.6 U/L in biliary atresia and NH/INH cases respectively which is not statistically significant. The mean serum alkaline phosphatase was 1048.8±162.8 U/L in biliary atresia and 629.6±160.8 U/L in NH/INH cases. Gamma glutamyl transpeptidase (ãGT), prothrombin time or serum albumin level showed no significant difference between biliary atresia and NH/INH cases.

The sensitivity and specificity of ultrasonography were 87.5% and 97.7% respectively[14]. Ultrasonography was done in all infants with cholestatic jaundice. In biliary atresia cases gallbladder was found either small in size or absent or no contraction of gall bladder was seen even after meals. These findings were consistent with the findings of other studies[2]. Visualization of a normal gallbladder while fasting and contraction after meal virtually rules out biliary atresia cases. But the reverse is not always true[15]. Thus USG of hepatobiliary system both before and after food may be used as differentiating between biliary atresia and NH/ INH.

Though scintigraphy is a competent diagnostic tool, its availability limits its use. Moreover, high jaundice may prevent uptake and further limits its use as a diagnostic tool in the community. The hepatobiliary scintigraphy (HIDA scan) identifies diseased gallbladders and bile drainage problems. Mandana (2009) showed that the sensitivity and specificity of HIDA scan was 100% and 50% respectively. Hepatobiliary scintigraphy (using 99 technetium iminodiacetic acids) was done in only selected patients when ultrasonography findings were not consistent with clinical finding. Nine patients showed delayed uptake but normal excretion which is consistent with NH/INH whereas five infants showed normal uptake of the isotope but absent excretion into the biliary channels and intestine which was consistent with biliary atresia. Similar findings were observed by other authors[16].

Percutaneous liver biopsy is the most valuable procedure in the evaluation of neonatal hepatobiliary diseases and provides the most reliable discriminatory evidence. Biliary atresia is characterized by bile ductular proliferation, the presence of bile plugs and portal and

perilobular edema and fibrosis, with the intact basic hepatic lobular architechture . In biopsy, features of biliary atresia were found in 12 cases and features of early biliary cirrhosis in 4 infants. Ten patients showed features of idiopathic neonatal hepatitis. Karim and Kamal[11] reported biliary atresia in 6 of 19 patients and biliary cirrhosis in four infants. They found 8 patients with idiopathic neonatal hepatitis. These findings are consistent with the present study. Though this is the main differentiating diagnostic procedure, its use is in community is limited due to lack of expertise and facilities in the wider community.

Conclusion:

Biliary atresia was found to be the commonest cause of cholestasis in this study. Most of the children presented late though appearance of jaundice was before two weeks of life. Acholic stool and USG finding before and after food appears to be differentiating and may be used by the primary care giver in identifying the problem and early referral.

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Internal Podalic Version an Option for Developing Countries

SF NARGIS[a], R ARA[b], G ARA[C], J BEGUM[d]

Summary:

Objective: To study the role of internal podalic version (IVP) in the management of transverse lie with fetal demise in labour and to assess the success and outcome of this almost obsolete procedure in obstetrics. Materials and Methods: Quasi experimental study was done in Shaheed Ziaur Rahman Medical College and Mohammad Ali Hospital, Bogra from January 2001 to December 2006. Results: Within this period total 180 cases transverse lie with intrauterine fetal death (IUFD) were studed. All these case were undiagnosed transverse lie with IUFD in labour and admitted in the hospital through the emergency department. In 60 cases IPV and in 120 cases lower uterine segment caesarean section (LUCS) were done to deliver the dead fetus. All the cases were more than 37 weeks gestation. Need for per operative and post operative blood transfusion were only 03.33% in cases where IVP was done, and 41.66% in those who under went LUCS (P<.001). Hospital stay in IVP group was only 1.3 days where as 10 days for those who under went LUCS (P<.001). Regarding complications it was almost same in both groups. IVP was also very cost effective required some of taka 300 only where as for LUCS it required about 3000 taka. Conclusion: The success rate of IVP is good. It is cost effective, preserve the future fertility and prevent subsequent scar rupture where Antenatal care is poor.

Key words: Transverse lie, Dead foetus, internal podalic version

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Introduction:

Version is defined as a shift in the position of the fetus inside of the uterus, either occurring naturally (spontaneous version) or as performed by a doctor to facilitate delivery. Internal version is an ancient procedure and it was extensively practiced by Hippocrates, who recommended internal cephalic version for all presentations other than head[1]. Aetius , Celsus and others at different times pointed out the fallacies of Hippocratic teaching and emphasized the advantage of internal podalilc version (IPV). IPV continued to be in favour till the sixteen century. In modern obstetrics caesarean section is the method of choice for the delivery of babies in transverse lie. The role of IPV followed by breech extraction is only limited to mal presentation or abnormal lie of the second twin.

In the case of singleton transverse lie it is contra indicated with a live fetus with intact membranes [1, 2].

a. Dr. Shamim Fatema Nargis, Associate Professor, Obstetrics and Gynaecology, SSMC.

b. Dr. Rawshan Ara, Assistant professor, Obstetrics and Gynaecology, SSMC.

c. Dr. Gulshan Ara, Associate Professor, Obstetrics and Gynaecology, SSMC.

d. Dr. Jahanara Begum, Associate Professor, Obstetrics and Gynaecology, SSMC.

Address of Correspondence: Dr Shamim Fatema Nargis, Associate Professor, Obstetrics and Gynaecology, SSMC.

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In the developing countries the maternal mortality rate (MMR) is very high and now in Bangladesh still it is 1.9/1000 live birth. Ninty percent deliveries are conducted at home and only 15% deliveries are conducted by Skilled Birth Attendants (SBA). Eighty percent maternal death occurred after home delivery[3].

Rupture uterus is a serious obstetric emergency with high maternal and perinatal mortality rate. The risk of uterine rupture is more in the presence of scarred uterus, lack of adequate antenatal care, low socioeconomic status and unsupervised labour at home. Performing a caesarean section for a dead, previable baby will lead to a scarred uterus increasing the risk of rupture in subsequent pregnancy.

Internal version is always podalic version and is completed with the extraction of the fetus. Before performing internal podalic version cervix must be fully dilated and liquor amni should be adequate for the intra uterine foetal manipulation. Patient should be deeply anaesthetized and in lithotomy position. Full surgical asepsis should be taken. If the podalic pole of the foetus is on the left side of the mother the right hand is to be introduced and vice versa. The hand is to be introduced in a cone shaped manner, it is than pushed up in to the uterine cavity keeping the back of the hand against the uterine wall until the hand reaches the podalic pole. The hand is to pass up to the breech and then along the thigh until a foot is grasped. The identification of the foot is

done by palpation of the heel and bring down the leg by a steady traction. While the leg is brought down the cephalic pole is pushed up by using the external hand. After one leg is brought down there is no difficulty in deliver the other leg by breech extraction.

IPV is much maligned to produce sepsis, postpartum haemorrage and uterine rupture causes high fetal and maternal morbity and mortality rates itself. In selected cases and in experienced hands IPV is comparatively safe in the developing countries and contribute to reducing the maternal mortality and morbidity by avoiding caesarean section and subsequent uterine rupture[4, 5].

Methodology:

It is a quasi experimental comparative trial done at Shaheed Ziaur Rahman Medical College and Mohammad Ali Hospital, Bogra from January 2001 to December 2006. The patients with transverse lie with intra uterine death at term admitted during this period were included in the study. In one group IPV was done and in other group caesarean was done to deliver the dead foetus. The mode of delivery either by IPV or by LUCS was recorded. LUCS was done in cases of impacted shoulder, impending rupture, failed attempt at version, and early sign of maternal shock due to obstructed labour or chorioamnionitis, possibility of intra uterine manipulation. LUCS also done in absence of experienced surgeon to perform IPV. Need for blood transfusion, cost effectiveness and hospital stay in two groups were compared. Complications occurred during version or LUCS were noted. Socio demographic details, associated obstertric history and parity were recorded. Complications were measured in terms of Post Partum Haemorrahage(PPH), para urethral or vaginal tear, rupture uterus, extension of wound or broad ligament haematoma and wound infections.

Results:

During 2001 to 2006 years almost total 24,000 delivery has occurred at shaheed Ziaur Rahman Medical College and Mohammed Ali Hospital, which is a teaching hospital and tertiary referral centre. Rate of LUCS is about 45% in this hospital due to different indications. Total percentage of transverse lie admitted within this period is 0.97 %(233cases). IPV were done in 60 (25.75%) cases, LUCS were performed in 120 cases (51.5%) of transverse lie, 53 cases presented with rupture uterus where laparotomy was done. 6 cases of IPV were in twin gestation. None of them were booked cases. Most cases (86%) came from rural population. 74.24% cases were of more than 37 weeks gestation, and 16cases (6.8%) were between 28 to 32 week gestation. 34cases (14.59) were primigravida and the rest 199 (85.40%) were multigravida. 53% of the patients were within the age range of 21 to 30. The groups of patients where IPV (60) was done, 8 (13.33 %) cases were primigravida and 52 (86.6%) cases were multigravida. During the procedure of IPV following complications like cervical tears in two cases, in10 cases paraurethral and vaginal tears occurred. In 1 case rupture uterus occurred during version and subtotal hysterectomy was done. Three mothers developed mild post partum haemorrhage and managed conservatively. Only in 2 cases blood transfusion was required. Almost all these patients were discharged from the hospital on the next day of admission.

Table-I

Shows the demography of the patients admitted with transverse lie and management given to them.

Total no of obstetric pt admitted		24000	
Total no of transverse lie admitted		233	
Age range (in years) no	15- 20 40 (17 %)	20-30 123 (53%)	30-40 70(30%)
Parity no	Primigravida 34	Multigravida 199	
Gestational age no	28 – 32 weeks 16	32-36 weeks 44	>37 weeks 173
Management given no	IPV 60	LUCS 120	Laparotomy

In 30 cases, version were done by obstetrician of >5 years of experience, 15 cases of version were done by obstetricians of 3 to 5 years experiences and 15 cases were done by junior doctors less than 2 years of experience with the supervision's of the seniors. General anaesthesia was given in all cases.

LUCS done on 20 (16.6%) cases were primigravid and 100 (83.33) cases were multigravid. During the procedure of LUCS complications like, extension of the uterine incision, broad ligament haematoma and vertical tear in the lower segment involving the vault of the vagina had occurred in 4 cases. 25 cases need blood transfusion both per and post operatively. General anaesthesia or Regional block was given in all the cases. Post operative stitch infection had occurred in 50 cases due to mal handling at home, and all needs secondary suture. But no burst abdomen was found. Mean postoperative hospital stay were 10 days.

Table-II

Show the difference in demography of the patient in IPV & LUCS groups.

	IPV(60)	LUCS (120)	Level of significance
Mean age	26.59	28	NS
Mean parity	3.00	3.2	NS
Mean gestational age	38 weeks	38weeks	NS

NS - not significant

Table-III

Show the difference of variable in between the IPV and LUCS

	IPV(60)	LUCS (120)	Level of significance
SAB/GA given	100% cases	100% cases	
Blood transfusion	02 (3.3%)	50(41.66%)	***
Hospital stay	1.3 days	10 days	***
cost	300 taka	3000taka	***

NS: - not significant

Table-IV

Showing the pattern of complications in of IPV and LUCS in cases of transverse lie.

	IPV	LUCS	Level of significance
PPH (mild to moderate)	02 cases	05 cases	NS
Para urethral or vaginal tear	10	10	NS
Cervical tear	02	00	NS
Rupture uterus	01	00	NS
Extension of uterine wound or Broad ligament haematoma	00	04	NS
Stitch infections need secondary suture	00	50	***

NS: - not significant

Discussion:

Analyzing the cases of transverse lie which were managed by IPV, none of these 60 cases had any form of antenatal care. They admitted in the hospital with complications when they could not deliver at home after various hours of labour pain. Due to improper Antenatal care there is high incidence of undiagnosed malpresentation in labour, and moreover due to inadequate transport facilities there is delay in approaching at the tertiary health centre. These complicated, undiagnosed cases of transverse lie report to the hospital in advanced labour and by this time it is not always possible to salvage their babies and even some times these mothers also. During this study period total 24000 patients admitted in this hospital and the incidence of transverse lie is 0.97% which is higher than normal incidence because it is a tertiary referral hospital. In our study of total 233 case of transverse lie admitted, out of which 38.83 %(60) under gone IPV procedure, 51.50 % (120) had LUCS and 22.74 %(53) cases comes with ruptured uterus.

Amita mahedru, Onome Ogueh, Ketan Gajjar, Charu Rawat in their study performed IPV in 15.8% and LUCS in 73.5% of total transverse lie admissions. 10.7% patient presented with rupture uterus in their study. Demography shows that 83% patients came from rural population, 88% were multipara, none of them were booked case, and 51% cases were more than 37 weeks gestational age which is similar to our study[1]. We found that 86% patient comes from rural area, 74.25% patient had gestational age more than 37 weeks, and none of them had any Antenatal check up except immunization against tetanus. Patient with rupture uterus is more (53cases) in our series. Chauhan AR, et al shows on their study from 1986 to 1991 and 1992 to 1997 that the incidence of transverse lie admitted was same in both series (0.4% and 0.32 %) but the number of IPV performed in the 2nd series reduced to almost halves. Most the IPV performed on the patient age between 20 to 30 years of age and on second gravida same as our study[4, 6]. Dufour et al conducted a retrospective study of 35 cases of IPV followed by breech extraction of second twin. They found that Internal Version together with external version is the only alternative to LUCS allowing rapid delivery of the second twin. Maternal prognosis is excellent and foetal out come is good if contraindications of IPV are avoided[7]. Rabinovici et al conducted a prospective study for management of second nonvertex twin. 60 twin deliveries after the 35th gestational week with vertex-breech and vertex-transverse presentation were managed according to randomized protocol of vaginal or abdominal delivery. Of 21 patient vertex transverse presentations 12 were delivered by LUCS. They found there was a significantly higher incidence of febrile morbidity in LUCSgroup[8]. In our study regarding many complications following IPV and LUCS are similar but in case of LUCS stitch infections need secondary suture in 50 cases and this complication is statistically significantly higher ($p<0.001$). pre operative and post operative blood transfusion is needed only 3.33% in cases of IPV and 41.66% in cases of LUCS operation , which is statistically significant (<0,001).

Conclusions:

In earlier years internal podalic version was performed on all stillbirths, nonviable babies, second twin and in a few singleton pregnancies in which the babies were of low birth weight and were preterm. However with improved neonatal facilities today, low birth weight and preterm babies have a better chance of survival. This combined with increased safety of the caesarean section, there is a changing trend in the mode of delivery of transverse lie more in favour of LUCS. Above all with the decrease trend for IPV, most obstetricians have not even seen it being performed. So caesarean section is performed even on dead babies and in large number of second twins. But rural patients who do not have health care facilities, LUCS with the increase risk of placenta previa, rupture uterus in subsequent pregnancy is always a threat.

We believe that though caesarean section is certainly a better option for singleton viable babies in transverse lie, IPV has a role to play in the delivery of second twin. It may also be attempted in the delivery of nonviable and dead babies in the absence of contraindications, especially in developing countries.

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Correlation between Estimated Fetal Weight at Term by Ultrasonogram and Actual Birth Weight

NR SHAPLA[a], MA ALEEM[b], E JESMIN[c], H AHMED[d], YS LEPE[e]

Summary:

The estimation of foetal birth weight is an important factor in the management of high risk pregnancies. Estimated foetal weight is calculated in the standard routine antepartum evaluation of high risk pregnancies and deliveries. This prospective observational study was done at the Department of Obstetrics and Gynecology in Border Guard Hospital, Peelkhana, Dhaka over a period of 6 months from January 2012 to June 2012. The present study was carried out to compare the accuracy of actual and ultrasonographic estimation of foetal weight at term. Hundred pregnant women at different gestational age from 37 weeks to 40 weeks were selected by simple random sampling. Ultrasonography was done for determination of estimated foetal weight (EFW) at term by using Hadlock method and birth weight was measured just after delivery. Data analysis was done by percentage and paired 't' test. The age range of patients were 18-37 years with mean ±SD is 25.13±4.46. Among 100 study patients 33% were nuliparous and 67% were multiparous. The mean ±SD of gestational age and actual birth weight is 38.76±1.09 and 3.11±0.391 respectively. Ultrasound biometric data that includes mean ±SD biparietal diameter (BPD) in mm, abdominal circumference (AC) in mm and femur length (FL) in cm were 90.21±3.52, 327.67±20.75 and 7.45±1.43 respectively. Mean ±SD of estimated foetal weight (EFW) Kg was 2.97±0.53. Actual birth weight is correlated with the estimated foetal weight and the result was not statistically significant (P >.05). Calculation of estimated fetal weight by ultrasonography is recommended to make decision about mode of delivery, so that an obstetrician can plan early in high risk cases.

Key words: Estimated foetal weight, Birth weight, Bipari et al diameter abdominal circumference.

(J Banagladesh Coll Phys Surg 2014; 32: 21-25)

Introduction:

Accurate estimation of fetal weight is of paramount importance in the management of labour and delivery.[1] It has long been established that birthweight is a major determinant of infant mortality in the first year of life and that mortality rates are more sensitive to brithweight than gestational age. Hence the importance attached to antenatal birthweight determination.[2]

The use of ultrasound for determination of fetal weight spans over three decades now, with varied attempts at the use of different biophysical parameters. Initial attempts to estimate fetal weight by ultrasound were made on the basis of measurements of individual single fetal parameters such as the Biparietal Diameter (BPD) or abdominal circumference (AC). Weight estimates obtained by these parameters were found to have high standard deviation up to 11.9%. Subsequent reports demonstrated that accuracy of the estimate was improved by the use of multiple fetal parameters. Further attempts to improve the predictive value of sonography in fetal weight estimation have resulted in the use of more parameters combined. Hadlock et al showed that using femur length (FL) in addition to head measurements and abdominal measurements significantly improved fetal weight estimation.[2]

This study is done to obtain an estimated fetal weight from the fetal parameters and to highlight the predictive value of this procedure by comparing the estimated fetal weight with the actual birth weight.

During the last decade estimated fetal weight has been incorporated into the standard routine antepartum evaluation of high risk pregnancies and deliveries. For instance, management of diabetic pregnancy, vaginal birth after a previous caesarean section and intrapartum management of fetuses presenting by the breech will be greatly influenced by estimated fetal weight.[1]

a. Dr Lt Col Nahid Reaz Shapla, Head of the Department ,Obs and Gynecology, Border Guard Hospital, Peelkhana, Dhaka.

b. Dr Lt Col Mohammad Abdul Aleem, Head of the department ,Anasthiology, Border Guard Hospital , Peelkhana , Dhaka .

c. Dr Lt Col Eva Jesmin, Head of the Department, Paediatrics, BG Hospital, Peelkhana.

d. Dr Hussain Ahmed, Junior consultant, Department of Radiology, BG Hospital Peelkhana, Dhaka .

e. Dr Yasmin Samad Lepe, Consultant, Obs and Gyne, BG Hospital, Dhaka.

Address of Correspondence: Lt Col Nahid Reaz Shapla, Gynaecologist, Border Guard Hospital, Peelkhana, Dhaka, Mobile: 01742181427.

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An acurate diagnosis of macrosomia for patients with gestational diabetes can reduce perinatal morbidity as it may assist the physician and staff in deciding the appropriate route of delivery, to prepare for shoulder distocia or to prevent a traumatic injury. Correct EFW values are also important when intrauterine growth is restricted and in preterm labour.[3]

The present study was undertaken to determine the accuracy of birth weight estimation by routine antepartum sonography at term.

Materials and Methods:

This prospective observational study was carried out in the Department of Obstetric and Gynecology in Border Guard Hospital, Peelkhana, Dhaka over a period of 6 months from January 2012 to June 2012. This study includes 100 pregnant women at term including obstetrical and medical complications (37-40 weeks), reliable date of last menstrual record, regular menstrual cycle, close correlation between menstrual age and clinical gestational age measurements, singleton pregnancy and live born infants without congenital malformation or hydrops. Women not at term, women with multiple pregnancies, advanced labour, antenatal diagnosis of congenital fetal malformation and intrauterine fetal death were excluded from this study.

Verbal consent from each patient was taken. After taking history with particular attention to aspects relevant to the study, clinical examination was done.

Once the diagnosis is confirmed the ultrasound examination was carried out by the same sonologist using 2D and 4D curvilinear probe by GE Voluson 730 Pro Scanner. Measurements were made with calibrated caliper on the machine on frozen images.

Biparietal Diameter (BPD) was made at the level of thalami from outer to inner table of the skull. Abdominal circumference was measured on the outer margin of the abdomen using internal calipers. Femur length measurements were taken by Hadlock method.

Estimated fetal weight is calculated by using standard Hadlock reference table that used biparietal diameter, abdominal circumference and femur length.

Birth weights were measured just after delivery.

Estimated fetal weight, patient demographic data and actual birth weight were recorded on data sheet that was kept separate from the patients chart. Student 't' test was done and level of significance was set at $P<0.05$ (5%).

Results:

One hundred pregnant women at term from 37 to 40 weeks gestational age were randomly selected.

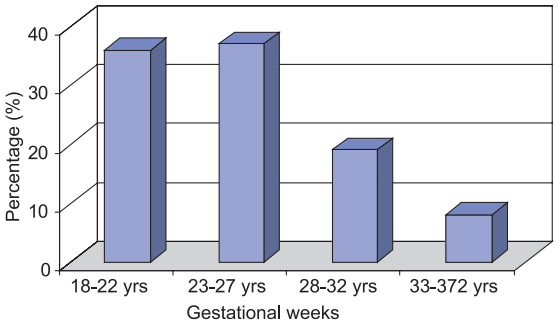

Fig- 1: *Age distribution of subjects.*

Figure 1 shows age range of the patients were between 18-37 years with a mean age of 25.13 ±4.46 years.

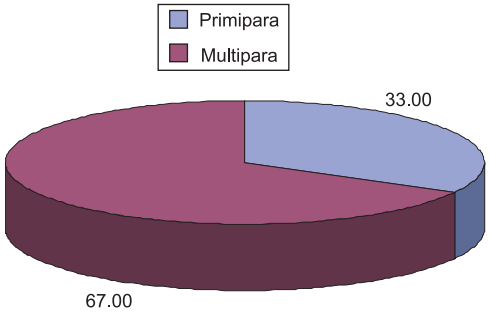

Fig- 2: *Distribution of parity.*

In figure 2 among 100 patients thirty three percent of gravidas were nulliparous and sixty seven percent were multiparous.

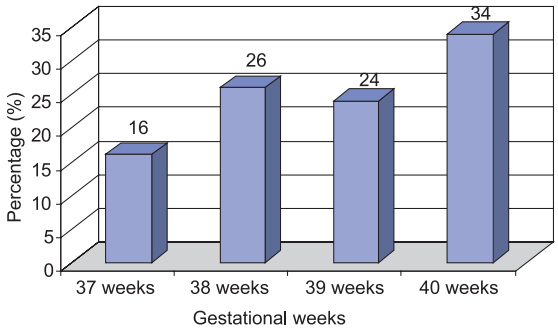

Fig- 3: *The distribution of patient by gestational age.*

In figure 3 among 100 patients 37 weeks pregnancy were of 16%, 38 weeks pregnancy 26%, 39 weeks pregnancy were of 24% and 40 weeks pregnancy were of 34%.

Table-I

Different ultrasonographic biometric data, EFW and birth weight at different gestational age.

Parameters (Mean±SD)	37 weeks	38 weeks	39 weeks	40 weeks
BPD (MM)	88.8±3.33	90.2±3.68	90.4±3.77	90.8±3.29
AC (MM)	323.0±22.15	334.8±18.35	328.3±22.22	324.0±20.12
FL (MM)	7.3±0.32	7.4±1.55	7.5±1.07	7.6±1.83
EFW (kg)	3.1±0.37	2.9±0.42	2.9±0.69	2.9±0.57
Birth weight (kg)	3.1±0.51	3.0±0.31	3.2±0.34	3.1±0.42

Table-II

Mean biparietal diameter (BPD), abdominal circumference (AC), femoral length (FL) and mean estimated foetal weight (EFW)

Characteristics	Minimum	Maximum	Mean	Std. Deviation
BPD (MM)	85	96	90.21	±3.52
AC (MM)	293	358	327.67	±20.75
FL (MM)	5.13	15.0	7.45	±1.43
EFW (kg)	1.049	4.20	2.97	±0.53

Out of 100 patients BPD were ranging 85mm to 96mm with mean 90.21±3.52, AC ranging 293mm to 358mm with mean ±SD of 327.67±20.75, FL ranging 5.13 to 15.0 with man ±SD of 7.45±1.43, EFW ranging from 1.049 Kg to 4.20 Kg with mean ±SD of 2.97±0.533

Table-III

Distribution of birth weight of newborn after delivery.

Birth weight	Number	Percentage
<2.5 Kg	14	14%
2.5-<4 Kg	73	73%
>4 Kg	13	13%

Among 100 patients 14% had birth weight <2.5 Kg, 73% had birth weight of 2.5-<4 Kg and 13% birth weight had >4 Kg

Table-IV

Mean ±SD of gestational age and birth weight after delivery.

Characteristics	Minimum	Maximum	Mean±SD
Gestational age (Weeks)	37	40	38.76±**1.09**
Birth weight (Kg)	2.0	4.5	3.11±**0.391**

Among 100 patients minimum gestational age was 37 weeks and maximum 40 weeks with mean ±SD of 38.76±1.09. Birth weight of newborn after delivery ranging from 2.10 to 4.5 Kg with mean ±SD 3.11±0.391.

Table-V

Discrepancy between mean birth weight and estimated fetal weight including P value.

Ultrasound Estimated fetal weight (EFW) Kg	Mean Birth Weight Kg	Mean true Birth weight -EFW Kg	P value
2.97±0.53	3.11±0.391	0.07	p>0.05

Among 100 patients mean EFW 2.97±0.53, mean birth weight after delivery 3.11±0.391 which shows no significant difference (P>0.05).

Discussion:

Accurate estimation of fetal weight has been shown to reduce parinatal mobidity and mortality associated with high risk pregnancy such intrauterine growth restriction and prematurity.[2] In present study the age range of patients was between 18-37 years with a mean age of 25.13±4.46 years. Akinula RA et al observed the age range of patients was between 16-41 years with a mean of 30.7 years.[2] Akinula S. S. et al showed that mean maternal age was 30.5±47 (range 22-41).[1] In this study 33% were primigravida and 67% were multiparous.

Akinula S. S. et al showed that 35% gravidas were multiparous and 60% were multiparous and which 5% were grand multiparous.[1] In this study mean gestational age ±SD of 38.76±1.09 with minimum gestational age was 37 weeks and maximum 40 weeks. Akinula S. S. et al observed that gestational age was 38.6±1.3 (range 37-42 weeks) which is almost similar to present study.[1] Akinula RA et al also observed almost the similar findings.[2] Juozas K. et al found in his study obtained from 5612 pregnant women. Fetal weight was estimated for each fetus using the formulas of Campbell and Wilkin, Shepard, 2 formulas of Hadlock and Merz. The result showed the best was Hadlock formula using 3 fetal biometry parameters. The lowest interclass correlation was found with Shepard formula.[4] In present study EFW is taken by ultrasound by Hadlock method. In this study EFW at 37 weeks 3.1±0.37 (Kg) and actual birth weight 3.1±0.51 (Kg) Akinula RA et al observed EFW at 37 weeks by Jadlock mathod mean ±SD 3290±123 and Actual Birth weight 3081±SD which is almost similar to present study.[2] In this study EFW at 38 weeks 2.9±0.42 and actual birth weight 3.0±0.31 (Kg) Akinula RA et al showed that EFW 3392±136 and Actual weight is 3338±385 which is almost similar with this present study.[2]

In this study at 39 weeks and 40 weeks EFW 2.9±0.69 and 2.9±0.57 respectively and Actual birth weight 3.2±0.34 and 3.1±4.2 Kg respectively which is also similar in the study observed by Aknula RA et al.[2]

After 36 weeks, the rate of weight gain steadily decreases in the normal fetus.[5] In our study after 37 wks to 40 wks mean EFW (Kg) shows steady decline from 3.1 ±. 0.37 to 2.9 ± 0.57.

In present study Mean Actual birth weight is 2.10 to 4.5 Kg with mean ±SD 3.11±0.391 which the mean EFW 2.97±0.533. So no significance difference between estimated fetal weight and actual birth weight.

Asrafganjooei T et al observed that the mean actual birth weight was 3329 (SD 443) g while the mean estimated fetal weights by ultrasound and clinical assessment were 3305 (SD 335) 3321 (SD 449).[3]

In one study done in Nigeria showed that clinical estimation of birth weight is as accurate as routine ultrasonographic estimation except in low birth weight babies. Therefore, when the clinical method suggests weight smaller than 2500 g, subsequent sonographic estimation is recommended to wield a better prediction and to further evaluation fetal well-being.[1] In contrast to this research we found that the accuracy of ultrasound for estimation of fetal weight is significant.

Hisham M. M. et al observed that the mean BPD measurement was 9.1±0.39 cm, mean AC was 34.1±3.0 cm and mean FL was 72±0.36 cm. the mean birth weight was 3418±541 gm. The correlation with actual term birth weight was highest with the formula of Shepard and Hadlock. The formula of Shepard and Hadlock had the minimum mean absolute percentage errors of 0.2 and 1.0 respectively.[6]

In present study mean BPD 90.21±3.52, mean AC 327.67±20.75, mean FL 7.45±1.43 and mean EFW 2.97±0.53. In this study the mean discrepancy between true birth weight and estimated fetal weight is 0.07. In present study, 14% babies were LBW with mean birth weight 2.97±0.53.

The positive predictive value of a sonographic estimate of fetal weight of < 2500 g is 87% for preterm fetuses, with a sensitivity of 90%, and the positive predictive value for a sonographic estimate of fetal weight < 1500 g is 86%, with a sensitivity of 93%.[7] A weight estimate above 4000 grams is associated with a 77% chance of macrosomia, and a weight above 4500 grams is associated with an 86% chance of macrosomia. The chance of macrosomia is only 16% when the weight estimate is less than 4000 grams.[8]

Conclusion

Low birth weight and excessive fetal weight at delivery both are associated with an increased risk of neonatal complications during labor and the puerperium. Birth weight has predicting value regarding survival and it is a useful parameter in predicting the susceptibility of diseases, future growth and development. In developing countries low birth weight is single most important factor that effects neonatal mortality and morbidity. Thus birth weight has largely been a subject of clinical and epidemiological importance and a target for public health intervention. Ultrasonography is an important tool for estimating fetal weight in uterus. The accuracy of ultrasound estimations of fetal weight before delivery in term pregnancies shows no significant difference with actual birth weight. So calculation of estimated fetal weight by ultrasonography is recommended to make decision about mode of delivery, so that an obstetrician

can plan early in high risk cases. Further large scale study is needed to establish the requirement of ultrasonography in each term pregnant women for estimation of fetal weight.

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REVIEW ARTICLE

Breast Feeding versus Formula Feeding and Diarrheal Diseases in Infants and Children- A Review

MUH BEGUM

Summary:

The World Health Organization (WHO) and the American Academy of Pediatrics (AAP) emphasize the value of breastfeeding for mothers as well as children. Both recommend exclusive breastfeeding for the first six months of life. Human breast milk is the healthiest form of milk for babies. Breastfeeding promotes health and helps to prevent diseases including diarrheal diseases. It contains all nutrients including antibodies (IgA),and lactoferrin, that potentially prevent infection and diarrhea in infants and children. Studies conducted in both developed and under developed nations have found that breast feeding is associated with significantly (upto 64%) less diarrheal disease and the protective effect of breast feeding does not persist beyond two months after breast feeding is stopped. On the other hand, formula fed infants are found an upto 80% increased in the risk of developing diarrhea compared to breast fed infants and there is significantly more diarrheal disease in formula fed infants. Infection may be attributable to contamination of bottles, teats, milk, and food in infants who are not exclusively breastfed. Exclusive breastfeeding for the first six months of life and there after complementary feedings while breastfeeding continues for up to two years of age or beyond, enthusiastic support and involvement from clinicians, obstetricians and pediatricians, are essential in "breastfeeding vs formula feeding" issue and to reduce incidence of diarrheal diseases in infants and children.

Keywords: World Health Organization (WHO), The American Academy of Pediatrics (AAP), sudden infant death syndrome (SIDS).

(J Banagladesh Coll Phys Surg 2014; 32: 26-30)

Introduction:

Breastfeeding and human milk are the normative standards for infant feeding and nutrition[1]. Breastfeeding results in improved infant and maternal health outcomes in both the industrialized and developing world. Breast milk is the best source of nutrition for the first 6 months of life. Breast milk contains all nutrients as well as antibodies especially Immunoglobulin A (Ig A) and protect baby from infections including diarrheal diseases. Though commercial infant formulas and cow's milk are considered nutritionally acceptable for infants, there is greatest risk of diarrheal diseases. Formulas and cow's milk as well as bottles, teats and utensil are attributable to contamination causing diarrheal diseases in infants who are not exclusively breastfed.

Address of Correspondence: Dr. Most. Umme Habiba Begum, Paediatric Department, Northern Private Medical College, Rangpur, Officer's Quarter no 103/6 (Sheuly), Rangpur Cantt, Mobile: 01191295626, 01712546647, e-mail: arafanis@yahoo.com

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Composition of breast milk and its anti-infective role:

Human breast milk is the healthiest form of milk for babies. It contains appropriate amounts of carbohydrate, protein, and fat, and provides digestive enzymes, minerals, vitamins, and hormones that infants require. Breast milk also contains antibodies from the mother that can help the baby resist infections. Colostrum and human milk contain an abundant amount of IgA[2]. It is secreted from the mammary and other exocrine glands during lactation. IgA prevents the attachment of bacteria and viruses to the gastrointestinal and other mucosal epithelium cells that would potentially cause infection and diarrhea. Human breastmilk also contains lactoferrin, being the main protein in human milk, This nourishment acts as a microbicidal agent killing bacteria and viruses[3].

Current recommendation on breastfeeding:

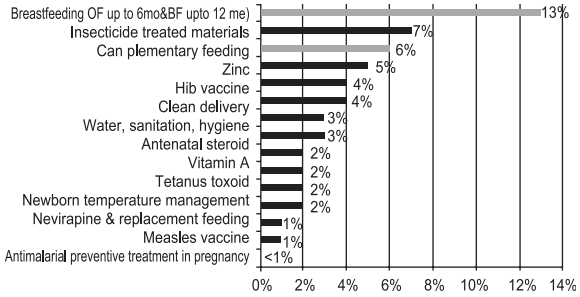
As a global public health recommendation (as per WHO and AAP), infants should be exclusively breastfed for the first six months of life to achieve optimal growth, development and health. Thereafter, to meet their evolving nutritional needs, infants should receive safe

and nutritionally adequate complementary foods while breastfeeding continues for up to two years of age or beyond[4]. Hospital practices that encourage successful breast-feeding include ante partum education and encouragement, immediate postpartum mother-infant contact with suckling, rooming-in arrangements, demand feeding, inclusion of fathers in prenatal breast-feeding education, and support from experienced women[5]. Though breastfeeding, including exclusive breastfeeding for the first 6 months of life, is widely advocated as "ideal" for babies and infants, less than 40% of infants below this age are currently exclusively breastfed worldwide[6].

Benefits of breastfeeding:

Scientific research has found numerous benefits of breastfeeding for the infant. American Academy of Pediatrics research has shown that breast feeding provides advantages with regard to general health, growth, and development. Not breastfeeding significantly increases risk for a large number of acute and chronic diseases including gastroenteritis, lower respiratory infection, ear infections, bacteremia, bacterial meningitis, botulism, urinary tract infection, and necrotizing enterocolitis. They state that there are a number of studies that show a possible protective effect of breast milk feeding against sudden infant death syndrome, insulin-dependent diabetes mellitus, Crohn's disease, ulcerative colitis, lymphoma, allergic diseases, digestive diseases, and a possible enhancement of cognitive development[7].

Breastfeeding is a cost effective way of feeding an infant, providing nourishment for a child at a small cost to the mother. Frequent and exclusive breastfeeding usually delays the return of fertility through lactational amenorrhea, though breastfeeding is an imperfect means of birth control. During breastfeeding beneficial hormones are released into the mother's body and the maternal bond can be strengthened. Breastfeeding is possible throughout pregnancy, but generally milk production will be reduced at some point[8]. The 2003 landmark Lancet Child Survival Series ranked the top 15 preventative child survival interventions for their effectiveness in preventing under-five mortality. Exclusive breastfeeding up to six months of age and breastfeeding up to 12 months was ranked number one, with complementary feeding starting at six months number three(Figure 1). These two interventions alone were estimated to prevent almost one-fifth of under-five mortality in developing countries[9].



(Source: Lancet Child Survival Series 2003)

Fig.-1: *Per cent of child deaths that could be prevented with 99% coverage of preventive interventions*

Formula feeding and health hazards:

Besides breast milk, infant formula is the only other milk product which is under the age of one year (as opposed to cow's milk, for instance). Supplementing with solid food in addition to breast milk or formula begins during weaning, and most babies begin supplementing about the time their first teeth appear, usually around the age of six months. Although cow's milk is the basis of almost all infant formula, plain cow's milk is unsuited for infants because of its high casein content and low whey content, which may put a strain on an infant's immature kidneys, and untreated cow's milk is not recommended before the age of 12 months. The infant intestine is not properly equipped to digest non-human milk, and this may often result in diarrhea, intestinal bleeding and malnutrition [10]. Since the early 1970s, industrial countries have witnessed a resurgence in breastfeeding among newborns and infants to 6 months of age. This upswing in breastfeeding has been accompanied by a deferment in the average age of introduction of other foods (such as cow's milk), resulting in increased use of both breastfeeding and infant formula between the ages of 3–12 months [11].

There are a number of formulas in vogue for infant feeding, such as raw milk formulas, evaporated milk formulas, commercial formulas, generic brand formulas, follow-on and toddler formulas[12] etc. But use of formulas including infant formula is associated with numerous increased health risks. Studies have found infants in developed countries who consume formula are at increased risk for acute gastroenteritis, otitis

media, severe lower respiratory tract infections, atopic dermatitis, asthma, obesity, type 1 and 2 diabetes, sudden infant death syndrome (SIDS), eczema, necrotizing enterocolitis and autism when compared to infants who are breastfed[13] .

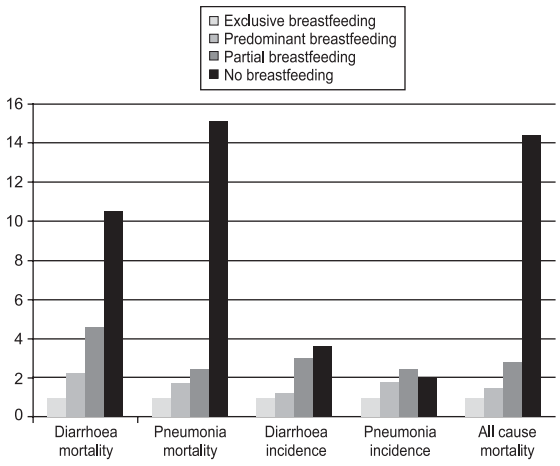
Breastfeeding and diarrhea:

Breast milk is the ideal food for an infant's first six months of life. In addition to providing ideal nourishment, breastfeeding provides infants with protection from many infections, including diarrheal diseases[14]. Infants are at greatest risk of diarrheal disease when foods other than breast milk are given. Breastfeeding frequency should be maintained for a year or more after adding food to the infant diet to reduce the risk of diarrhea that may have serious consequences to health, nutritional status, and survival. When breastfeeding stops, infants are exposed to food-borne germs and lose the protection of breast milk's anti-infective properties such as lactoferrin, IgA, oligosaccharides. When infants do experience diarrhea, severe dehydration can occur quickly. Continued breastfeeding during diarrhea, as well as increased feeding after an episode, significantly reduces risk of dehydration, mitigates loss of weight, and promotes increased weight gain. Breastfeeding can also reduce the severity, duration, and negative nutritional consequences of diarrhea[14].

Discussion:

Breast-fed children, compared with the bottle-fed ones, have a lower incidence of acute gastroenteritis due to the presence of several antiinfective factors in human milk[15]. Human milk contains oligosaccharides which prevent infections related to some common pathogenic bacteria. In a study, Coppa GV, et al. has shown that fractions of oligosaccharides of human milk contains, acidic oligosaccharides, neutral high-molecular-weight oligosaccharides, and neutral low-molecular-weight oligosaccharides. The acidic fraction had an antiadhesive effect on pathogenic strains of enteropathogenic Escherichia coli serotype O119, Vibrio cholerae, and Salmonella; the neutral high-molecular-weight fraction significantly inhibited the adhesion of E. coli O119 and V. cholerae; the neutral low-molecular-weight fraction was effective toward E. coli O119 and S. fyris. Human milk oligosaccharides inhibit the adhesion to epithelial cells not only of common pathogens like E. coli but also for V. cholerae and S. fyris. Consequently, oligosaccharides are one of the important defensive factors contained in human milk against acute diarrheal infections of breast-fed infants[15].

The risk of morbidity and mortality from suboptimum breastfeeding in young children has been documented in observational studies. Robert E Black et. al. observed in a random effects meta-analysis[16] that increased risk of cause-specific morbidity and mortality in relation to four patterns of breastfeeding in children younger than 6 months (exclusive—ie, nothing but breastmilk; predominant—only water or teats in addition to breast-milk; partial—other liquids or solids in addition to breast-milk; and not breastfeeding), and two patterns (breastfeeding or not) in children aged 6—23 months. In the first 6 months of life, the relative risks were increased for each of the three patterns that were compared with the reference pattern—ie, exclusive breastfeeding, for diarrhea and pneumonia morbidity and mortality (Figure 2). The relative risks were significant for predominant breastfeeding for all-cause mortality and pneumonia incidence, and there were similar, but not significant, point estimates for diarrhea and pneumonia mortality and diarrhea incidence. Compared with exclusive breastfeeding, partial breastfeeding had moderately higher relative risks than predominant breastfeeding, and not breastfeeding had very high relative risks. In infants aged 6—23 months there was a statistically raised risk of not breastfeeding for all-cause mortality and diarrhea incidence, but there was no significant raised risk for other outcomes[16].



(Source: Programming Guide: Infant and Young Child Feeding; the IYCF Unit, UNICEF, June 2012)

Fig.-2: *Relative risk of not breastfeeding for infections and mortality compared to exclusive breastfeeding from 0-5 months*

Case-control study of diarrheal disease among 304 infants (167 cases and 137 controls) cases presenting to 34 general practices in England showed breast feeding was associated with significantly less diarrheal disease. Associations were striking even in infants aged e" 6 months. They did not vary by social class, but were greater in those living in rented council accommodation and in more crowded households. The effect of receiving no breast milk was stronger in more deprived areas than in less deprived areas. The effect of not receiving exclusive breast milk was stronger in more deprived areas than in less deprived areas. In formula fed infants, there was significantly more diarrheal disease in those not sterilizing bottles/teats with steam or chemicals. The protective effect of breast feeding did not persist beyond two months after breast feeding had stopped. Breast feeding protects against diarrheal disease in infants in England although the degree of protection may vary across infants and wear off after breast feeding cessation. Education about the benefits of breast feeding and the risks of inadequate sterilization should be targeted at carers in deprived areas or households[17]. A large volume on the evidence for the many benefits of breastfeeding in industrialized countries has been compiled. It shows a 64% lowered risk for gastrointestinal tract infections[18].

In the longitudinal analysis of infants in the United States conducted by Scariati, Grumer-Strawn, and Fein, researchers found an 80% increase in the risk of developing diarrhea in the formula fed children compared to breast fed infants. Investigators also found a "dose response effect that a small but steady increase in the risk of developing diarrhea as the amount of breast milk an infant received decreased"[19].

An episode of diarrhea was significantly less likely to last for six or more days if an infant was breastfed for three or more months shown in a study by Baker D et al. in "Inequality in infant morbidity: causes and consequences in England in the 1990s"[20].

The type of milk consumed before start of diarrhea episode was strongly associated with dehydration. Compared with infants exclusively breastfed, bottle-fed infants were at higher risk (odds ratio for cow's milk = 6.0, for formula milk = 6.9). Compared with those still breastfeeding, children who stopped in the previous two months were more likely to develop dehydrating diarrhea observed in a study by Fuchs SC et al. in "Case-control study of risk of dehydrating diarrhea in infants in vulnerable period after full weaning"[21].

In the first year of life the incidence of diarrheal illness among breastfed infants was half that of formula-fed infants observed by Dewey KG et al. in "Differences in morbidity between breast-fed and formula-fed infants." study[22]. Children less than 12 months of age had a lower incidence of acute diarrheal disease during the months they were being breastfed than children that were fed with formula during the same period shown by Lerman,Y. et al.in "Epidemiology of acute diarrheal diseases in children in a high standard of living settlement in Israel"[22]. Strictly formula-fed children had an incidence of diarrhea over three times that of strictly breast-fed infants and twice that of breast-fed and supplemental fed children observed by Long KZ et al. "Proportional hazards analysis of diarrhea due to enterotoxigenic Escherichia coli and breastfeeding in a cohort of urban Mexican children"[22]. In the study Blake PA, et al. "Pathogen-specific risk factors and protective factors for acute diarrheal disease in urban Brazilian infants" of 500 Brazilian infants d" 12 months old with diarrhea and 500 age-matched controls, breast-feeding infants <6 months old (OR, 0.3) and boiling household drinking water (OR, 0.4) were protective. Breast-feeding was protective against enteropathogenic Escherichia coli infections (OR, 0.1). The addition to the breast-milk diet of even water, teas, and other nonnutritive liquids doubled or tripled the likelihood of diarrhea. Supplementation of breast-feeding with additional nutritive foods or liquids further increased significantly the risk of diarrhea observed by Popkin BM et al. in "Breast-feeding and diarrheal morbidity" study[22].

Whereas breast-fed infants have less severe diarrhea when breast milk is continued rather than interrupted, infants and children fed with non-human milks tend to have more severe illness than those receiving milk-free or lactose-limited formulas or milk-cereal mixtures, observed in study by Jorge L Lembcke et al." Effect of milk-containing diets on the severity and duration of childhood diarrhea"[23].

Conclusion:

Breast milk cannot be duplicated by any artificial means. It is unique in its composition and function. Breastfeeding has been shown to reduce morbidity and mortality rates due to diarrheal diseases in infants and

children worldwide. Different studies have consistently reported a decrease in the incidence of diarrhea in breastfed infants and children. Moreover by breast feeding, mothers are providing a natural form of nourishment, reducing long term healthcare cost, eliminating formula cost, and nurturing a bond with her child. As clinicians, recommendations should be made to soon-to-be parents regarding the importance of breast milk during routine obstetric and pediatric visits. Enthusiastic support and involvement of the pediatricians in the promotion and practice of breastfeeding is essential to reduce incidence of diarrheal diseases and thereby the achievement of optimal infant and child health, growth, and development.

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Porokeratosis with an Invasive Squamous Cell Carcinoma: A Case Report and Review of Literature

M NESSA[a], L KHONDKER[b], MA BARI[c], MSI KHAN[d], M RAHMAN[e], SY MONI[f]

Summary:

Porokeratosis is a clonal disorder of epidermal keratinization, which is characterized by hyperkeratotic papules or plaques that are surrounded by a thread-like elevated border. The histopathologic hallmark of porokeratosis is the cornoid lamella, which is a thin column of parakeratosis that overlies a thin or absent granular layer and that corresponds to the raised, hyperkeratotic border. Porokeratosis has five clinical types and malignant degeneration has been described in all forms of porokeratosis. We report a forty five year old farmer with a large plaque in chest for 30 years and multiple nodules within the large plaque for 2 years. A section of skin from margin of the plaque reveals histopathological features of porokeratosis and section from nodules reveals an invasive squamous cell carcinoma. To the best of our knowledge, this is the first reported case of porokeratosis transformed to squamous cell carcinoma on Bangladesh.

Key words: Porokeratosis, invasive squamous cell carcinoma.

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Introduction:

Porokeratosis is a clonal disorder of keratinization characterized by one or more atrophic patches surrounded by a clinically and histologically distinct ridge-like border called 2 cornoid lamella2 . [1,2] Five major clinical variants are recognized – classical or plaque type porokeratosis of Mibelli (PM), disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis (LP), porokeratosis palmaris et plantaris disseminata (PPPD) and punctate porokeratosis (PP).[1,3] Distinction between these types is made purely on clinical criteria.[4] There are reports of more than one type of porokeratosis developing in the same patient.[5] The most common form of porokeratosis is DSAP, typical lesions have symmetric distribution and usually affects sun-exposed areas. [6] The etiology of porokeratosis remains unclear. Malignant degeneration has been reported in all forms of porokeratosis, with a reported incidence of 7.5 to 11 percent.[7] The occurrence of malignancies in porokeratotic lesions is clinical evidence of the pre-cancerous nature of this disease.[8] Over-expression of p53 in lesional epidermis may play a role in malignant transformation. In addition, DNA flow cytometry analysis has demonstrated abnormal DNA ploidy in the lesional epidermis of patients with different types of porokeratosis. The most commonly associated malignant conditions with porokeratosis are Bowen disease, squamous-cell carcinoma (SCC), and, rarely, basal cell carcinoma. The greatest risk is attributed to large lesions, long duration, and linear-type lesions.[9] Squamous cell carcinoma arising in the classic type of porokeratosis of Mibelli is well-documented, but there are only a few reports of squamous cell carcinoma in DSAP. [10] There is no definitive treatment for Porokeratosis. Porokeratosis does not usually need treatment, but in some cases, treatment is necessary due to potential for progression to a malignancy and for

a. Dr Moriom Nessa, Assistant Professor, Dept of Dermatology and Venereology, Islami Bank Medical College Hospital, Nawdapara, Rajshahi.

b. Dr. Lubna Khondker, Assistant Professor, Department of Dermatology and Venereology, Bangabandhu Sheikh Mujib Medical University (BSMMU), Shahbag, Dhaka, Bangladesh.

c. Dr. M A Bari MS (Urology), Rajshahi Medical College Hospital, Rajshahi.

d. Lt Col (Dr) Md Shirajul Islam Khan, Graded Specialist in Dermatology and Venereology, Combined Military Hospital (CMH), Dhaka cantonment, Dhaka.

e. Dr. Moksedur Rahman, Junior Consultant, Department of Dermatology and Venereology, Upazila Health Complex, Rajshahi.

f. Dr. Sabiha Yeasmin Moni, Rajshahi Medical College Hospital, Rajshahi.

Address of Correspondence: Dr. Lubna Khondker, Assistant Professor of Dermatology, Room-209(B), 2nd floor, Block-B, Bangabandhu Sheikh Mujib Medical University (BSMMU), Shahbag, Dhaka. Mobile- 01552370429; E-mail: lubna_ derma @yahoo.com

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cosmetic purposes.[11] Attempts to treat with topical, systemic, and intralesional glucocorticoids; topical 5-fluorouracil; phototherapy; and keratolytics have yielded only marginal benefits. Surgical modalities, such as cryotherapy, shave excision, curettage, linear excision, and dermabrasion have been used to treat small lesions with variable success. It has been suggested that oral retinoids may reduce the risk of malignant transformation. Relapses are common within weeks to months after cessation of oral retinoid therapy. [12]

Case report:

A 45 year old farmer from Porsha, Nowgaon reported to department of Dermatology and venereology, Islami Bank Medical College Hospital, Rajshahi, with a large plaque in chest for 30 years and multiple nodules within the large plaque for 2 years. According to the statement of the patient, he was alright 30 years back, and then he first developed a papule in the left chest just under the left nipple. Then it spreads slowly towards periphery and become depressed centrally with some scaly lesion in the centre, later it becomes serpiginous large well defined plaque, surrounded by a keratotic wall. This wall is grayish and surmounted by a tiny groove along its summit. Now left sided chest, left abdomen and left back is occupied by a large plaque. First, it was symptom less and he gives no history of burning, itching or pain. In the meantime he went to some local doctors and he took some medicine from doctors but failed to cure. He is a farmer and he works in the sunlight in the field. He gave history of trauma in the patch 6 years back, at the time of catching fish. After 28 years of that lesion, he noticed multiple nodules growing within the plaque, some are small and one is large nodule. Large nodule is situated below the left nipple, at the site of origin. The nodule is everted, cauliflower shaped with some exudation within centre. Now patient gives history of itching and burning. Patient is non-diabetic and normo-tensive. He gives no history of taking chemotherapy, PUVA, organ transplantation or no abnormality is detected in the mucous membrane, hair and nail. Nothing contributory history of drug, family or personal history. On examination, the plaque is surrounded by a keratotic border. The nodule is large, everted, cauliflower shaped and exudative in nature.

All the hematological(TC, DC,ESR, Hb%) and biochemical(Random blood sugar, serum ALT, serum creatinine) parameter was within normal limit. From two sites of skin, biopsy material was taken, one from margin of the plaque and another from large nodule and sent for histopathological examination. A section of skin from margin of the plaque reveals mild hyperkeratosis and a shallow depression containing columns of parakeratotic cells. Epidermis below this area shows dyskeratotic changes and the dermis reveals a moderate infiltration of chronic inflammatory cells. These features are confirmatory of porokeratosis, on histo-pathologically, though classical or plaque type porokeratosis variety on clinically. Section from nodules reveals an invasive squamous cell carcinoma. The tumor is well differentiated. It has invaded the subcutis. The deep margin shows presence of tumor (Grade-I). Patient was treated by topical steroid and retinoid and advised for excision of nodule.

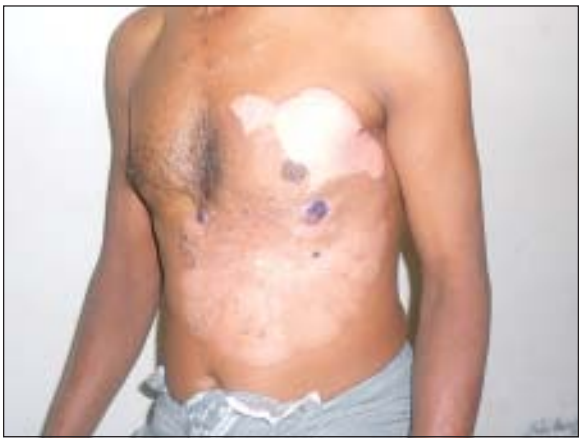

Fig.-1: *Photograph of the patient with a large well defined plaque, surrounded by a keratotic wall in the chest and multiple nodules within the large plaque.*

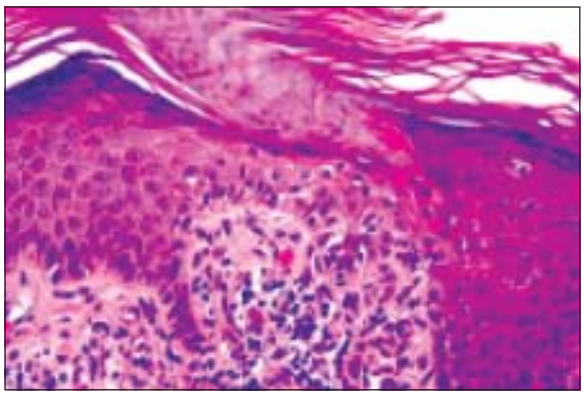

Fig.-2: *Photograph of histopathological finding from margin of the plaque showing - cornoid lamella of porokeratosis.*

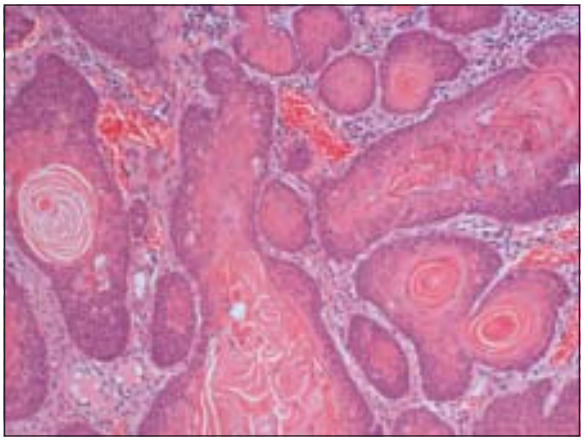

Fig.-3: *Photograph of histopathological finding from large nodule showing Squamous cell carcinoma (SCC)*

Discussion:

Sengupta Sujata et al reported a case of 40-year-old farmer (case 1) with asymptomatic skin lesions over the extremities, abdomen, groins, palms and soles for the past ten years. This case was reported in the department of Dermatology, R.K.M Seva Pratisthan and Vivekananda Institute of Medical Sciences, Kolkata, India. Since the last 6 months, he noted the appearance of multiple raised mildly itchy but painless growths over the pre-existing lesions in the groins. His family history was not suggestive of a similar disease. He denied an exposure to radiation and there was no history of high-risk sexual practices. No evidence of immuno-suppression was apparent. On examination, numerous small well-defined hyperpigmented plaques, some of which were more than 10 cm in diameter, were seen. These giant lesions were present in the abdomen, thighs and soles. The involvement of the palms and soles was prominent. Individual plaques had an atrophic center with a prominent border that was traversed by a thread-like groove. In the inguinal region on both sides there were multiple erythematous papulonodular lesions of varying sizes, which bled on manipulation. The largest of them was 5cm x 3cm x 2cm. A few inguinal lymph nodes palpable were less than 0.5cm in size, soft to firm, not tender and mobile. No other lymph node was palpable. The rest of the cutaneous and systemic examination was normal. Routine blood tests, biochemical examinations, X-ray of the chest were normal. VDRL and ELISA for HIV were nonreactive. Histology from the edge of a flat lesion showed a parakeratotic column (cornoid lamella) in the epidermal invagination with absence of the underlying granular layer. A non-specific perivascular infiltrate of chronic inflammatory cells was seen in the dermis. These findings were consistent with porokeratosis. Fine needle aspiration and cytology (FNAC) from two contralateral inguinal nodules showed atypical cells and histopathology revealed features of a well-differentiated SCC in both. Ultrasonography of the abdomen showed no organomegaly, lymph node enlargement or any evidence of metastasis. A CT-guided FNAC of the inguinal and femoral lymph nodes showed reactive hyperplasia without any cellular atypia. The diagnosis of multicentric SCC arising from PPPD and giant porokeratosis was reached.[13]

Nilendu Sarma et al reported a 51-year-old male (case 2) and his 23-year-old son with multiple, well-defined, annular oval, polycyclic or irregular shaped lesions with raised margins in the Department of Dermatology, NRS Medical College, Kolkata, India. All the lesions were dark brown and were widely distributed both in the exposed as well as the covered areas. They were dry, palpable, and the margins were elevated compared with the central area. Along the margin, there was a prominent deep furrow. Most of the lesions measured 4-6 cm in their longest dimension and were slightly larger in size in the father. The father had some additional features. Many lesions, even smaller ones, showed a tendency towards development of gross hypertrophy in the margin with well formed horns in some. The central part of the lesions showed clinically normal skin in most lesions. Some lesions also appeared warty with a prominent break on the entire surface. In the father, there was a large ulcer (8 x 9 cm) over both the buttock and perianal areas with predominant lateral extensions towards the patient's left side (at 3 to 6 o' clock) and towards the patient's right side (9 to11 o' clock). The ulcer was surrounded by typical lesions with hyperkeratotic margin. The ulcer involved the anal margin exposing the mucosal layer that appeared mildly prolapsed. There were also multiple lesions scattered over the buttocks; their sizes were 3 to 6 cm and mostly oval in shape. Considering the clinical presentation, they diagnosed these cases as disseminated Mibelli's plaque type porokeratosis with multiple horns and an ulcer in the father. The father's grandmother, his both sons (including one who attended), and daughter had similar diseases. His father was asymptomatic until 33 years of

age when he died of unknown cause. His mother did not have the same disease. All who had the disease started to develop the lesions during the age of 12-15 years. Histological examination of the lesion was done in the father from two sites; one from the margin of a well-defined hyperkeratotic lesion over the foot and another from the ulcer in the perianal region. Histology from the margin of clinically typical porokeratosis over the foot showed a column of severe hyperkeratosis and epidermal invagination filled with parakeratosis (coronoid lamella). The subepidermal stroma showed mild infiltration of lymphocytes. This histomorphological feature was consistent with our clinical diagnosis of porokeratosis.A histopathological examination of the ulcer in the perianal region showed atypical squamous cells in clusters and sheets with surface ulceration. The squamous cells showed moderate pleomorphism, hyperchromatic nuclei, and prominent nucleoli. Brisk mitosis and focal necrosis were observed. Keratin pearl and evidence of intracytoplasmic keratin were noted. On the basis of these histological features, a diagnosis of keratinizing squamous cell carcinoma was made. The tumor involved the anal canal reaching up to the squamo-columnar junction in its upper part. The anal sphincteric muscles were also infiltrated by the tumor. A histological examination of a skin lesion on the back from the son also showed features consistent with porokeratosis of Mibelli. Finally, the cases were confirmed as familial porokeratosis of Mibelli's type with malignant transformation to squamous cell carcinoma over the buttock involving the anus in the father. Unfortunately, the patients were lost during the work-up period even before any therapeutic intervention could be made.[14]

A 73-year-old woman (case 3) was referred to the Disorders of Keratinization Clinic at Bellevue Hospital Center 27 years ago with scaly lesions that began on her soles and progressively spread to her palms, legs, thighs, upper extremities, trunk, neck, and face. Her mucous membranes were spared. The papules on the palms and soles became increasingly hyperkeratotic and confluent into thick plaques. The palmoplantar lesions are painful due to pressure and interfere with ambulation and manual dexterity. Some of the other lesions are pruritic, especially in the summer and on exposure to heat. The patient has a history of atrial fibrillation that is treated with warfarin, dofetilide, and propranolol. She also has hyperlipidemia that is treated with atorvastatin. After an evaluation of bone density for retinoid-associated osteoporosis, treatment with alendronate was initiated. She has no known allergies to medications or family history of dermatologic diseases. She worked in a factory sewing cases for cosmetic products for more than 20 years although she retired in 2001. Histopathologic examination of a biopsy speciman from a lesion on her right upper thigh at the time of presentation confirmed the suspected clinical diagnosis. A biopsy specimen obtained in 2002 disclosed the same diagnosis. She was initially treated with isotretinoin daily, which resulted in a decrease in the number of lesions on the body and in the hyperkeratosis of the papules on the palms and soles. In 1985, the medication was changed to etretinate and this medication was even more effective in decreasing the number of lesions and the palmoplantar hyperkeratosis. In 1998, after removal of etretinate from the market, acitretin was initiated at a dose of 25 mg daily, with similar clinical efficacy as the parent compound. Attempts to discontinue or decrease systemic retinoids have resulted in pain and pruritus that interfere with her quality of life. Conversely, attempts to increase the dose of acitretin have been limited by both cheilitis and systemic adverse effects, which include blackouts. Numerous adjunctive treatments to diminish hyperkeratosis have been tried; these include salicylic acid 6 percent gel, salicylic acid in 10 to 40 percent concentrations, and urea 20 to 50 percent with and without occlusion. These agents have facilitated debridement of the palms and soles, which has been routinely performed by the patient and periodically in the office. Prolonged treatments with topical imiquimod and fluorouracil neither produced improvement nor prevented formation of new lesions. Topical tretinoin had to be discontinued because of erythema and pruritus. Destructive modalities, which included 90 percent trichloracetic acid, cryotherapy, and electrodesiccation have been successful in treating individual lesions. Physical examination reveals innumerable 2-3 mm; grey, annular papules with well-demarcated, slightly raised, hyperkeratotic borders covered her face, chest, back, abdomen, and extremities. Lesions were present in both sun-exposed and sun-protected areas. On the palms and soles, there were numerous, dicrete, non-punctate lesions as well as a confluence of papules, some with elevated borders. Her mucous membranes were spared.

A complete blood count with differential analysis and comprehensive metabolic panel were normal. Histopathological findings reveal focal thinning of the epidermis, with loss of the granular layer, and a discrete column of parakeratosis. [15]

A 62-year-old male (case 4) presented with pruritic eruptions on sun-exposed portions of both forearms that had gradually increased in number over a period of 5 years. The lesions were exacerbated during the summer months. Along with these lesions, an erythematous, irregular, marginated, scaly, crusted plaque had developed on the right forearm 3 years earlier. There was no significant medical or family history.An irregular, marginated, erythematous plaque and multiple, brown, atrophic macules surrounded by well-demarcated, raised ridges on the right forearm.On physical examination, a 2×3 cm erythematous, irregular, marginated, scaly, crusted plaque was noted on the right forearm. In addition, the patient had numerous annular, brown, atrophic, and symmetric macules surrounded by well-demarcated, raised ridges on extensor aspects of both forearms, which are characteristics of DSAP. Complete blood count, as well as liver and kidney function tests were all within normal limits. A skin biopsy specimen of the multiple, brown, annular lesions showed histologic changes of typical DSAP. There was a cornoid lamella composed of a column of parakeratosis with underlying hypogranulosis and perivascular lymphocytic infiltrations in the dermis localized beneath the cornoid lamella. A skin biopsy obtained from the erythematous plaque on the right forearm showed dysregulated keratinocytes with hyperchromatic, atypical nuclei, consistent with squamous cell carcinoma. A cornoid lamella was observed in the lesion of the squamous cell carcinoma. Biopsy specimen obtained from the erythematous plaque on the right arm. In epidermis, acanthosis and dysregulated keratinocytes with hyperchromatic, atypical nuclei are observed. A cornoid lamella composed of a column of parakeratosis is seen in the lesion. Positron emission tomography-computed tomography revealed no evidence of distant metastasis. The squamous cell carcinoma was treated by total excision and split-thickness skin graft and radiotherapy. The patient is currently being treated with topical sunscreens. [16]

Researchers in the department of Dermatology in the University of Tokushima, Japan reported that they have shown p53 over-expression immuno-histochemically in 14 of 17 porokeratotic specimens obtained from 14 lesions of nine cases, and in all six specimens of squamous cell carcinoma (SCC) arising on porokeratotic lesions of two cases. We screened mutations in exons 5 to 10 of the p53 gene in all these specimens by polymerase chain reaction-single strand conformation polymorphism analysis. Mutations of the p53 gene were detected in two of the six SCCs but not in any of the 17 porokeratotic specimens. These two mutations were C to T transitions at codons 146 and 175 in exon 5, which were a nonsense mutation at a dipyrimidine site and a missense mutation at a CG site, respectively. To our knowledge, neither of these mutations has been identified in skin cancers before. Our observations indicate that mutations of the p53 gene are not the major molecular etiology for porokeratosis, but are related to its skin carcinogenesis, and that p53 overexpression in porokeratosis is not due to p53 gene mutations. [17]

Christine Liang reported a 22-year-old man(case 5), presented to the Dermatology Clinic at Bellevue Hospital Center in July, 2008, with a two-year history of warts on the shaft of his penis, thighs, scrotum, and perianal area. An external biopsy report brought in by the patient had been interpreted as condyloma acuminata. The patient was treated at an outside clinic with podophyllin and cryotherapy and noted improvement of the perianal lesions but limited improvement of the lesions on the penis and scrotum. Past medical history was noncontributory. The patient was sexually active with both men and women and reported a history of unprotected sex. He denied a prior history of sexually transmitted diseases. The patient took no medications. A punch biopsy was obtained from a lesion on the scrotum. Physical Examination reveals that on the dorsal shaft of the penis were multiple, small, annular plaques with a thin, threadlike border. Numerous, arcuate, erythematous plaques with borders consisting of small erythematous papules were noted on the scrotum. On the left inguinal fold were multiple verrucous papules and plaques. The perianal area was clear. A rapid plasma reagin test was non-reactive. Urine DNA amplification testing for gonorrhea and chlamydia were negative. Within a hyperplastic epidermis there is a dell with hypogranulosis, dyskeratosis, and an overlying column of parakeratosis, which is consistent with a cornoid lamella. Beneath this is a band-like lymphohistiocytic infiltrate. [18]

A 25 year male (case 6) reported with skin lesions of 12 years duration. The lesions started as small keratotic papules over his perineum which spreaded peripherally very slowly and eventually formed well defined plaques surrounded by irregular keratotic border with atrophic center. The lesions gradually spreaded to penis, upper

thighs, feet and a few lesions scattered over trunk, face and scalp. There was no history of contact with chemicals or any medications. The lesions were skin colored, hairless and of variable sizes. All relevant investigations were within normal limits. Fungal microscopy, fungal culture, montoux test, STS were negative. Skin biopsy for histopathological examination revealed features of consistence with porokeratosis Mibelli. Genetic analysis could not be done due to lack of facilities. From this case 1, their study was able to trace 9 other members in successive generations of his family with porokeratosis. The case 1(propositus) inherited the disease from his maternal grandfather. The individuals affected in 3 successive generations are 10 in numbers. Of the 10 cases, 9 were examined personally and had histological evidence of porokeratosis; the rest one was reliably reported by close relatives to have the disease. None of the cases eventually developed any cutaneous malignancy. Among the ten affected patients 5 were male and 5 were female. In generation- I, one out of two; In generation- II, two out of five; In generation- III, seven out of fourteen family members were affected. The age of affected members ranged from 18 to 17 years and the age of disease onset was between 12 to 17 years. In that study, authors could not find out any malignant degeneration in any case. Cases in 3 successive generations suggest that porokeratosis is a genetic disorder and not a simple dyskeratosis. [19]

Conclusion:

A case of malignant transformation from classical or plaque type porokeratosis is reported here. Malignant change has been reported in nearly all types of porokeratosis. A review of malignant transformation found that these changes were more frequent on non-exposed skin, in large porokeratosis lesions, and in patients who previously received radiation therapy. Therefore, long-term follow-up for porokeratosis lesions is reasonable and long-term follow-up studies are needed.

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Cholecystocutaneous Fistula following Drainage of Parietal Abscess: A Rare Case Report

MAA AMIN[a], ME ULLAH[b], N SHABNAM[c]

Summary:

Spontaneous perforation of gallbladder as a complication of biliary stones may lead to cholecystocutaneous abscess or fistula. Here we report a case of cholecystocutaneous fistula in a 50-year-old diabetic female patient who presented with a chronic discharging sinus on right upper abdomen with recurrent abscess formation which failed to heal despite repeated attempts at incision drainage and debridement. After evaluation the tract was explored and was found to be communicating with the fundus of the gall bladder. The whole fistula tract was excised along with cholecystectomy.

(J Banagladesh Coll Phys Surg 2014; 32: 37-40)

Introduction:

Most cholecystocutaneous fistulae are postoperative complications of liver and biliary tract surgery or trauma. External biliary fistulae rarely occur spontaneously as a result of intrahepatic abscess, necrosis or perforation of the gallbladder, or other inflammatory process involving the biliary tree[1]. Spontaneous cholecystocutaneous fistulae are often the result of neglected biliary tract disease. This is rarely observed today because of the early diagnosis and management made possible by ultrasonography, broad-spectrum antibiotics, and effective surgical management of biliary tract disease[2]. The pathophysiology of this condition has been associated with increased pressure in the gallbladder, secondary to biliary obstruction. Ultrasonography or computed tomography and fistulography play an important role in the diagnosis of cholecystocutaneous fistula. The treatment of such fistula requires elective cholecystectomy with excision of the fistula tract and drainage of any associated abscess with adequate antibiotic coverage.

a. Dr. Md. Abdullah-Al-Amin, Professor & Head, Department of Surgery & MISC, BIRDEM & Ibrahim Medical College, Dhaka, Bangladesh.

b. Dr. Mahmud Ekram Ullah, Assistant Professor, Dept of Surgery & MISC, BIRDEM & Ibrahim Medical College, Dhaka, Bangladesh.

c. Dr. Nilufar Shabnam, Registrar, Dept of Surgery & MISC, BIRDEM & Ibrahim Medical College, Dhaka, Bangladesh.

Address of Correspondence: Dr. Md. Abdullah -Al –Amin, Professor & Head of the Department, Surgery & MISC, BIRDEM & Ibrahim Medical College, Dhaka, Bangladesh, Email: alamin.abdullah00@gmail.com

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Case report:

Mrs. X, a 50 year old diabetic lady hailing from Comilla got admitted in BIRDEM Hospital on 22/11/2010 with a chronic discharging sinus on right upper abdomen for 8 months. She had a history of drainage of parietal abscess over the same area 9 months back followed by secondary closure of the wound. But she repeatedly developed wound infection afterwards and the wound never really healed completely. She noticed recurrent seropurulent discharge through the central part of the scar. She had no history of nausea, vomiting, anorexia, fatty food intolerance, and fever or weight loss.

On examination, a drawn in scar (8-10 cm) was noted in the right hypochondrium with a small opening in the centre. On gentle compression, seropurulent discharge came out through the opening. Surrounding tissue was indurated but non-tender and local temperature was not raised. Abdominal and other systemic examination were unremarkable.

Our first clinical impression was tubercular sinus. The wound swab microscopy did not reveal any AFB. However, exploration of the tract was needed anyway and we planned to send the tract / tissue scrap for histopathological examination. MT or other serological tests were deemed unnecessary in this case.

Her other baseline hematological and biochemical investigations were all within normal limit. Wound Swab for C/S reported no growth. USG of abdomen demonstrated irregular hypoechoic area in muscle layer suggesting collection. However , it also showed sludge in gall bladder lumen (Fig-I). Sinogram revealed a linear tract and the contrast medium passed into a cavity (Fig-II).

Under G/A the sinus tract was explored by excising the previous scar and some seropurulent collection in the

subcutaneous space was drained. On further exploration, it was found to be communicating with an intraabdominal viscus. So the incision was extended and deepened as right subcostal incision through which abdominal cavity was explored. The fistula tract was found connected to the fundus of the gall bladder. Gall bladder was found adherent to the parietes, filled with pus and a stone was impacted in the cystic duct. The whole fistula tract was excised along with cholecystectomy. Her post-operative period was uneventful.

Histopathology of gall bladder reported as acute on chronic cholecystitis and tissue from fistula tract showed infiltration of mostly chronic and also acute inflammatory cells with in the wall of the tract. No granuloma or malignancy was seen. So our final diagnosis was chronic empyema of the gall bladder with spontaneous cholecystocutaneous fistula.

Fig-1: *USG showing irregular hypoechoic area in muscle layer*

Fig-2: *Contrast X-ray delineating the tract and a cavity*

Discussion:

Spontaneous cholecystocutaneous abscess or fistula is an uncommon complication of gallbladder disease that has been known since the time of Thilesius in 1670. Courvoisier documented 499 cases of gallbladder perforation in the late 19th century; 169 of these cases formed cutaneous tracts[3]. Over the past century, there have been fewer accounts of this problem because of prompt and safe management of biliary tract disease. In their review in 1949, Henry and Orr found 36 cases of external biliary fistulae reported after 1890[3]. In 2004, Vasanth A, Siddiqui A and O'Donnell K. have reported 20 cases over 'past 50 years'[2].

External biliary fistulae or abscesses rarely occur spontaneously following necrosis or perforation of the gallbladder, or other inflammatory process involving the biliary tree. In the past, external drainage of the abscess and antibiotics were used for sepsis control. However, this approach has been associated with biliary fistula formation. Most cholecystocutaneous abscesses or fistulae are however iatrogenic or traumatic[4].

In a review in 1994, Kaminsky reported on the frequency of biliary fistula. In this series, cholecystocutaneous abscesses or fistulae accounted for only 2% of all the cases[4]. Generally, the fistulae or abscesses appear in the right upper quadrant, although other locations such as the epigastrium, umbilical area, right groin and even the gluteal region have also been described[5].

Spontaneous cholecystocutaneous fistulas are almost always a result of neglected biliary tract disease[2]. Patients with this complication usually do not have classic symptoms normally associated with cholecystitis in their history, since this would have brought such a patient to seek medical attention sooner. Their symptoms tend to be more non-specific and often masked by co-existent disease. Symptoms of underlying disease may be ignored by the patient or may be masked by some underlying neuropathy[6]. This is consistent with the presentation in our case. Apart from the complaints of repeated local pain and swelling during episodes of infection, she did not have the classic symptoms of cholecystitis.

The patients are usually women over the age of 60[7]. However, cases have been documented in patients as

young as 24 years old. The fistula usually presents itself as an enlarging mass before spontaneous rupture. In some cases, it may be associated with symptoms of upper abdominal colic, dyspepsia, jaundice, or weight loss[6].

The pathophysiology of this condition has been associated with increased pressure in the gallbladder, secondary to cystic duct obstruction, most commonly due to calculi and rarely due to gallbladder carcinoma[5]. The increase in intraluminal pressure leads to impairment of the blood flow to and lymph drainage from the gallbladder, thus causing mural necrosis and perforation. Subacute perforation results in an abscess formation around the gallbladder with the formation of an internal or external biliary fistula. These fistulas, as presented in this case, frequently arise from the fundus of the gallbladder. The state preceding spontaneous rupture has been termed "empyema necessitatis" by Nayman[8]. This term essentially describes a "burrowing abscess" of the abdominal wall as a result of gallbladder inflammation.

The external opening of the fistula can be confused with a pyogenic granuloma, infected epidermal inclusion cyst, or metastatic carcinoma[6]. The discharge from the fistula may be purulent and mucoid if the cystic duct is obstructed by a stone as was found in our case. In cases where the cystic duct remains patent, discharge may be bilious. Intraoperatively the gallbladder usually appears small, contracted, chronically inflamed, and adherent to the parietes[7].

Our patient had no particular symptoms attributable to gall bladder disease. Even though her gall bladder was filled with pus, she was otherwise asymptomatic. Chronic empyema is such a condition that is sometimes seen particularly in diabetic elderly population where clinical manifestation of inflammatory response is subdued. Advancing age, diabetic autonomic neuropathy and sub-optimal use of antibiotics may be an explanation. In their large study comprising 1392 patients having gallbladder disease, Thornton J R and his colleagues showed that empyema of the gallbladder is sometimes chronic, painless, and afebrile. [9]

Imaging plays an important role in the diagnosis of this complication. Before fistula formation, the abscess can be diagnosed via ultrasonography, with findings that include a sonolucent mass with echogenic material adjacent to the anterior abdominal wall[10]. Sino/fistulogram, allows visualization of its origin and course[5]. Ultrasonography and CT imaging can also help in the diagnosis of this complication.

Management of cholecystocutaneous fistula should initially include control of any acute inflammatory process. This can be done by incision and drainage of the abscess if any, followed by wound cultures and appropriate antibiotic therapy. However, no bacteria could be isolated in our case. Surgically, the fistula tract can be laid open, with removal of any gallstones present in poor risk patient[11]. Spontaneous healing of cholecystocutaneous fistula has been reported in some cases[3]. However standard treatment is elective cholecystectomy along with excision of the fistula tract and drainage of any associated abscess.

Conclusion:

This case report demonstrates that maintaining a high degree of suspicion of this rare entity is helpful during surgery. The possibility of cholecystocutaneous fistula should be considered in any patient who has a discharging sinus in the right upper abdominal or chest wall. Early cholecystectomy and excision of the fistula tract can reduce morbidity from this complication. However, continued advances in noninvasive investigations, and widespread practice of elective and emergency cholecystectomy will eventually put an end to this exceedingly rare complication of common gallbladder disease.

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Successful Outcome of Pregnancy of a Sub-fertile Woman with Multiple Fibroids and Placenta Praevia and Accreta

M SULTANA[a], P AKHTER[b], AK SAHA[c], A AKHTER[d]

Summary:

Fibroids, the commonest benign pelvic tumor, have a common association with subfertility and increasing maternal age. Placenta praevia and fibroid (if submucous) has an association with placenta accreta. Here we report the case of an elderly primigravida with history of prolong subfertility, admitted with 37 weeks pregnancy with multiple fibroids and central placenta praevia with focal increta. Even with all these complicating factors, with the advent of better uterotonic drugs, conservative management could be considered successfully now-a-days.

Key words: Caesarean myomectomy, Fibroid, Placenta praevia/accreta, Postpartum haemorrhage.

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Introduction:

Fibroids are the commonest of all pelvic tumours, being present in 20 percent of women in reproductive age group. They are frequently multiple and more common in nulliparous and relatively infertile women[1,2]. Most fibroids are asymptomatic during pregnancy and are diagnosed on routine ultrasound examination[2]. Approximately 10% of pregnant patients with fibroids have complications related to or caused by the fibroids. These can be: first trimester loss, compression effect from myomas on the mother as well as on the fetus, pain (red degeneration), premature labour, premature rupture of the membranes, malpresentation, placenta praevia, retained placenta, postpartum hemorrhage and puerperal sepsis[3,4].

On the other hand, placenta accreta is a condition in which all or part of the placenta is abnormally adherent to the uterine wall[5,6,7]. Incidence varies from 1 in 2000 to 1 in 7000 deliveries[8]. Because of partial or total absence of the decidua basalis and imperfect development of fibrinoid layer, placental villi are attached to the myometrium in placenta accreta (75-80% of cases). With placenta increta, villi invade into the myometrium (15-17% of cases). With placenta percreta, villi penetrate the full thickness of the myometrium (5-7% of cases)[5,6,7,8,9]. This variant can lead to the placenta attaching to other organs such as the urinary bladder and rectum[9]. Here we report a case of term pregnancy with multiple fibroid with central placenta praevia, in which case focal placenta increta diagnosed peroperatively. During management of such a case, all preparation including sufficient blood should be in hand. Proper counseling should include consent for hysterectomy. Obstetric and anaesthetic expertise is the most important part for management.

Case Report:

A 37 years old, primigravid lady, hailing from Chowdhury Bary, Narayanganj, housewife of a lower socio-economic class, got herself admitted in 200 Bedded Hospital, Narayanganj on 21st April, 2012 with 37 weeks pregnancy with multiple fibroids and central placenta praevia. The patient is the 2nd wife in her 2nd married life and her present husband has 2 children from his previous wife. She had history of prolong subfertility of about 20 years (including both 1st and 2nd marriage).

For the first time, she came to the Gynae OPD, 200 Bedded Hospital, Narayanganj five months back on 28 November, 2011 with lower abdominal pain for three days which was severe for one day and slight per vaginal bleeding for two days. An urgent USG was done which shows about 16 weeks pregnancy with central placenta praevia with multiple fibroids within the body of uterus (largest one measuring 8.5cm X 7.4cm). Her pregnancy was first diagnosed at that time by USG and her EDD

a. Dr. Masuda Sultana, Junior Consultant (Gynae), Upazila Health Complex, Meghna, Comilla.

b. Dr. Parul Akhter, Junior Consultant (Gynae), 200 Bedded Hospital, Narayanganj.

c. Dr. Asim Kumar Saha, Junior Consultant (Gynae), Upazila Health Complex, Beanibazar, Sylhet.

d. Dr. Ayesha Akhter, Senior Consultant (Gynae), Sadar Hospital, Norsingdi.

Address of Correspondence: Dr. Masuda sultana, Flat- C4, H/N-128, R/N- 9/A, Dhanmondi R/A, Dhaka, Mob: 01713451898, 01678131094, Email: masudasultana22@gmail.com

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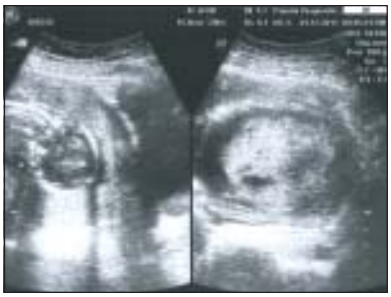
was on 9th May 2012. She gave history of repeated per vaginal bleeding in early pregnancy which she confused with irregular menstruation. She was admitted on that day, treated conservatively and discharged with treatment after 7 days. Thereafter, her pregnancy period was uneventful and she was on regular antenatal check-up in Gynae OPD of 200 Bedded Hospital, Narayanganj.

At 37 weeks, she developed mild lower abdominal pain and slight per vaginal bleeding for several hours. Once again she got herself admitted urgently. After proper counseling and keeping two units of fresh blood and two donors ready, caesarean section was done on the same day. A healthy male baby weighing 3 kg was delivered. Placenta was found in the lower segment, completely covering the internal os from the posterior uterine surface. It could not be separated by controlled cord traction. It was unusually adherent in the upper part, from where complete manual removal was possible but focal adherence of the placenta into the myometrium was found in the posterior part of the lower segment, from where it was removed incompletely by piece meal. A small submucous fibroid measuring about 1.5cm X 1.5cm was found there which was removed by finger dissection. Mattress suture was given in that part and uterine incision was closed. Four other myomas (intramural) of different sizes were found in the anterior wall and fundus of uterus, largest one measuring about 8cm X 7cm. They were removed by two planned incisions and incisions were closed by haemostatic sutures. Keeping a drain in situ abdomen was closed in layers. Two units of fresh blood were transfused peroperatively. Within two hours, the patient developed PPH and was managed by oxytocin, ergometrin and misoprostol. Intrauterine balloon tamponade was administered at that time and again two units of fresh blood were transfused. Rest of the post partum period was uneventful. Patient was discharged on 10th post partum day. Both the mother and the baby were healthy during discharge.



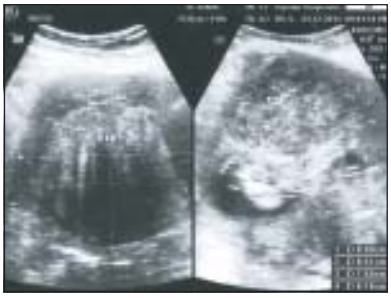


Fig. 1 & 2: *Ultrasonography showing multiple fibroid with 16 weeks of pregnancy in this patient.*

Discussion:

Uterine fibroids are seen in 1.6-4% of pregnancies. With increasing age of obstetric patients, more cases are being encountered during pregnancy[2,4]. Ultrasonography is helpful not only in diagnosing the presence of fibroid with pregnancy, but also in finding out size, number and site of fibroids and their relation to the placenta[10-]. Uterine fibroid during pregnancy is usually managed expectantly and surgically removal is generally delayed until after pregnancy[10]. Pedunculated fibroids can easily be removed and hemostasis secured at the time of cesarean delivery without endangering life of the mother. Yet, myomectomy at the time of cesarean section was practically absent from the obstetric literature until the last two decades[11].

Myomectomy at the time of cesarean delivery has traditionally been discouraged because of high risk of intra- and post-operative complications, such as uterine atony, uncontrollable hemorrhage and postpartum sepsis[12,13,14,15,16]. The medical literature has reported an increase in myomectomy during cesarean section in the past two decades[17]. This procedure is not always hazardous and it can be performed without significant complications by experienced obstetricians[12]. Rather, at delivery and puerperium a high incidence of hysterectomy for postpartum hemorrhage and postpartum sepsis was observed in some literature in which myoma was not removed[13]. There are some reports of huge, symptomatic leiomyomas successfully managed by myomectomy in second trimester[17,18]. This seemed to lead to an improvement in pregnancy outcome in carefully selected patients[18]. However, myomectomy performed during pregnancy remains a rarity[17]. In our

case, although myomas were symtomatic at 16 weeks of pregnancy, the patient got relieved with conservative management.

Placenta accreta occurs when there is a defect in the decidua basalis, allowing the anchoring villi to adhere to the myometrium. The frequency of abnormal placentation has increased 10-fold over the last 20 years and is now observed in 9.3% of women with placenta praevia or in 1 per 533 deliveries[19]. It is a life-threatening problem that is rising in incidence throughout the world. The increased risk of this problem in women with placenta praevia and one or more prior cesarean deliveries is well established. Other risk factors include previous uterine curettage, submucous uterine myomas, previous myomectomy, Asherman syndrome, maternal age older than 35 years, smoking and elevated á-fetoprotein levels[5,9,19]. Problems associated with delivery of placenta vary appreciably, depending on the site of implantation, depth of myometrial penetration and number of lobules involved[5]. Focal or partial involvement may be manifested as difficulty in establishing a cleavage plane during manual removal of placenta. Removal of a totally adherent placenta is difficult. Persistent efforts to remove a totally adherent placenta manually results in an even more blood loss and nearly always ends with caesarean hysterectomy[5,8]. There are a few case reports of patients with placenta accreta where caesarean hysterectomy was done due to intraoperative bleeding. In a recent study of 315 patients who required hysterectomy for antepartum or intrapartum bleeding, 38% had placenta praevia, and 68% of them also had placenta accreta, increta, or percreta[19]. Fortunately, our case has focal placenta increta where there was an option for conservative management. Placenta was removed manually incompletely by piecemeal from the adherent part. We found a submucous fibroid which was removed by finger dissection. To achieve haemostasis, mattress suture was given in that part. With the use of high dose of oxytocin and misoprostol peroperatively, myomectomy was not very hazardous in this case.

Conclusion:

With the increasing incidence of both placenta accreta with praevia and fibroid in pregnancy, obstetrician should carefully select the cases where conservative management could be tried. Per-operative haemorrhage is the prime indicator for abandoning the desire for conservation of uterus. Post-partum heamorrhage and puerperal sepsis again may end in hysterectomy in such a patient. In spite of that, myomectomy during cesarean section can be a safe and effective procedure in carefully selected patients in experienced hands. Successful management is a team work which requires co-operation among obstetricians, anaesthetists, physicians and haematologists.

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Diaphragmatic Hernia with Atypical Presentation - A Case Series

SMA SHAHID[a], MN ALI[b], M AHMED[c], SS ISLAM[d], SB HOSSAIN[e]

Summary:

Congenital Diaphragmatic Hernia is one of the most challenging diagnosis faced by pediatric surgeons. From the time of its first anatomic description more than 300 years ago, CDH has carried a high mortality rate. We aimed to review patients who presented with hernia of diaphragm during the last six months. In this retrospective study, the medical records of three patients treated for diaphragmatic hernias who were admitted to Rajshahi Medical College Hospital between July 2012 and December 2012 were analyzed. Three patients with age of 45 days to 7 years were included in the study. Male to female ratio was 1:2. All patients had left-sided diaphragmatic hernia. Chest X-ray was obtained from all patients which was diagnostic. One patient needed thoracotomy incision. No patient required mesh repair. The mean hospitalization time was 14 days. There was no postoperative death. Diaphragmatic hernia is an uncommon and challenging situation for the surgeon. Prompt diagnosis and treatment prevent serious morbidity and mortality associated with complications such as gangrene and perforation of herniated organ.

Keywords: Diaphragmatic Hernia, Diaphragm, Gastric volvulus, colonic gangrene

(J Banagladesh Coll Phys Surg 2014; 32: 45-50)

Introduction:

Congenital diaphragmatic hernia (CDH) is a developmental defect of the diaphragm that allows abdominal viscera to herniate into the chest.[1]

The embryologic development of the diaphragm involves multiple, complex

cellular and tissue interactions. The fully developed diaphragm is derived from four distinct components: (1) the anterior central tendon forms from the septum transversum, (2) the dorsolateral portions form from the pleuroperitoneal

membranes, (3) the dorsal crura evolve from the esophageal mesentery, and (4) the muscular portion of the diaphragm develops from the thoracic intercostal muscle groups. The precursors of diaphragmatic structure begin to form during the fourth week of gestation with the appearance of the peritoneal fold from the lateral mesenchymal tissue. At the same time, the septum transversum forms from the inferior portion of the pericardial cavity. The septum transversum serves to separate the thoracic from the abdominal cavities and eventually forms the central tendinous area of the fully developed diaphragm[1-2].

Anatomically, the right side closes before the left. Muscularization of the diaphragm appears to develop from the innermost muscle layer of the thoracic cavity, although it has been proposed that the posthepatic mesenchymal plate is a possible source of muscular tissue. Posterolaterally, at the junction of the lumbar and costal muscle groups, the fibrous lumbocostal trigone remains as a small remnant of the pleuroperitoneal membrane and relies on the fusion of the two muscle groups in the final stages of development for its strength. Delay or failure of muscular fusion leaves this area weak, perhaps predisposing to herniation[3]

Bochdalek first described this area of the posterolateral diaphragm in 1848, and it is for this reason that the most common site for CDH bears his name[1-3] Other types include Morgagni hernia, diaphragm eventration and central tendon defects of the diaphragm. Bochdalek hernia is the result of a congenital defect in the posterior costal part of the diaphragm in the region of the 10th and 11th ribs, which allows free communication between the thoracic and abdominal cavities [4] .

a. Dr. Shah Md. Ahsan Shahid, Assistant Prof., Department of Paediatric Surgery, Rajshahi Medical College Hospital, Rajshahi.

b. Dr. Md. Nawshad Ali, Department of Paediatric Surgery, Rajshahi Medical College Hospital, Rajshahi.

c. Prof. Dr. Mushtaque Ahmed, Department of Paediatric Surgery, Rajshahi Medical College Hospital, Rajshahi.

d. Dr. Sayed Sirajul Islam, Department of Paediatric Surgery, Rajshahi Medical College Hospital, Rajshahi.

e. Dr.S.Belal Hossain, Department of Paediatric Surgery, Rajshahi Medical College Hospital, Rajshahi.

Address of Correspondence: Dr. Shah Md. Ahsan Shahid, Asst. Professor, Department of Paediatric Surgery. Rajshahi Medical College Hospital.Rajshahi-6000, Mobile: +8801711277286, Email: smashahid293@gmail.com

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CDH is estimated to occur in 1 out of 2000 to 5000 births. Females appear to be more commonly afflicted than males. Affected neonates usually present in the first few hours of life with respiratory distress that may be mild or so severe as to be incompatible with life [5,6]. With the advent of antenatal diagnosis and improvement of neonatal care, survival has improved but there still remains significant risk of death and complications in infants with CDH[7]. The overall mortality is still high at most centers[8].The major cause of death is due to two complications: pulmonary hypoplasia and pulmonary hypertension[9]. Experts disagree on the relative importance of these two conditions, with some focusing on hypoplasia, others on hypertension[10].

Because herniation occurs during a critical period of lung development when bronchial and pulmonary artery branching occurs, lung compression by the herniated bowel results in pulmonary hypoplasia. With increasing severity of lung compression, there is a corresponding decrease in the bronchial branching resulting in a reduction of generations of bronchi and lung tissue. In addition, arterial branching is reduced and there is muscular hyperplasia of the pulmonary arterial tree [10-11].

Current ideas view the defect to be an inherent abnormality of lungs which results in a secondary abnormality of the diaphragm or as a failure of the diaphragm to separate the Pleuroperitoneal canal into thorax and abdomen before the midgut returns from umbilicus.The resultant abnormality leads to disordered lung growth. Both lungs are affected,th ipsilateral mo so than the contralateral. Affected infants are bon with a complex interface of pulmonary hypoplasia and pulmonary hypetension. Pulmonary hypoplasia can be severe enough to preclude life outside the womb, while successful management of pulmonary hypertension can lead to a fruitful life[12-14].In this report, we present three cases of diaphragmatic hernia with atypical presentation.

Objectives

Clinical presentation and management of three cases of congenital hernia of diaphragm were discussed to highlight challenges in diagnosis and handling of complications.

Patients and Methods

Three patients with the diagnosis of congenital diaphragmatic hernia treated in Rajshahi Medical College hospital between July 2012 and December 2012 were enrolled to this study. The mechanism, duration of complaints, clinical presentation, kind of visceral herniation, surgical repair, and outcome of the patients have been scrutinized. Diaphragmatic hernia repair and organ reduction were performed in all patients through abdominal approach except one,that required additional thoracic approach.

Case Report-1

A 7-years-old female child presented to our institution with complaints of abdominal pain, non passage of stool for five days, abdominal distension, frequent bilious vomiting, and respiratory distress for three days. The pain was progressive and colicky in nature and became more severe with no definite radiation or shifting. The vomiting was bilious and foul smelling and abdominal distension was progressive. Past medical history was significant and since birth she has been suffering from recurrent respiratory distress especially after running, playing, jogging and was relieved by taking rest. She also suffered from intermittent gastrointestinal symptoms like epigastric discomfort and postprandial colicky abdominal pain. She visited consultant pediatrician and received medication like antibiotics, nasal drops, antihistamines, gastroprokinetcs and antiflatulent for recurrent respiratory tract infections.

Her perinatal history was insignificant and she was delivered by caesarian section and her birth weight was 3.25 kilograms. On general physical examination the patient was restless,dehydrated,febrile, tachypnic with granting respiration. she was moderately anemic, moderately cyanosed and nonicteric. Abdominal examination revealed tense, moderately distended and tender abdomen. Tenderness was more marked over epigastric and left hypochondriac region where rebound tenderness was also present. Abdomen was resonant on percussion, absent bowel sound and no sign of ascites present. Digital rectal examination revealed normal findings. On chest examination asymmetry found between left and right hemithorax, the point of maximum cardiac pulsation and trachea shifted to right. Breath sound was normal on right but present only apex on left side. Laboratory investigations revealed neutrophilic leucocytosis and mild hypokalaemia. Abdominal and chest radiographs in erect postures revealed shifted trachea and mediastinum to right, gas distended bowel loops with air fluid levels in left hemithorax.

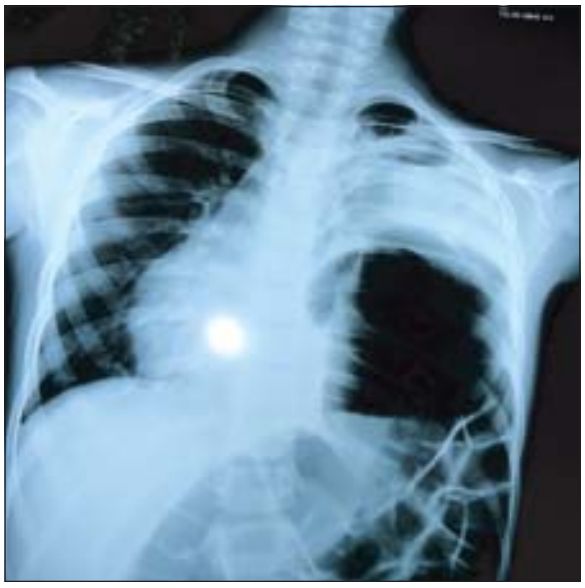

Fig.-1: *Chest radiograph showing ill-defined left dome of diaphragm and multiple air-fluid level in mid &lower zone of left hemithorax and mediastinal shift to right.*

The patient was provisionally diagnosed to have obstructed diaphragmatic hernia. After resuscitation emergency surgery was arranged. abdomen was approached through left subcostal incision. Moderate amount of hemorrhagic fluid found in abdominal cavity and small intestine, caecum, ascending and transverse colon were hugely distended. The left dome of diaphragm was highly elevated. The stomach, greater part of intestine and spleen with its hilum was found in the left hemi thorax. Part of transverse colon was protruded through an aperture of the diaphragm. The viscera were gently reduced from left hemithorax. The reduction of transverse colon was difficult as it found herniated through the diaphragmatic defect and finally it was irreducible. it required an additional thoracic approach and a left sided thoracotomy done. There was a defect on left dome of diaphragm which was about 5 cm in diameter. Herniated intrathoracic part of transverse colon was gangrenous.

After reduction of abdominal contents a tension free diaphragmatic repair was done with 1/0 prolene and the laxed part of diaphragm was plicated. Chest wall was closed, keeping a water seal drain in situ.

The gangrenous part of transverse colon was resected and a double barrel colostomy was performed on right hypochondrium. Abdomen was closed in layers. In immediate post operative period the patient needed ventilatory support in intensive care unit for 24 hours. Later she could maintain a satisfactory oxygen level and post operative chest radiographs revealed optimum lung expansion.

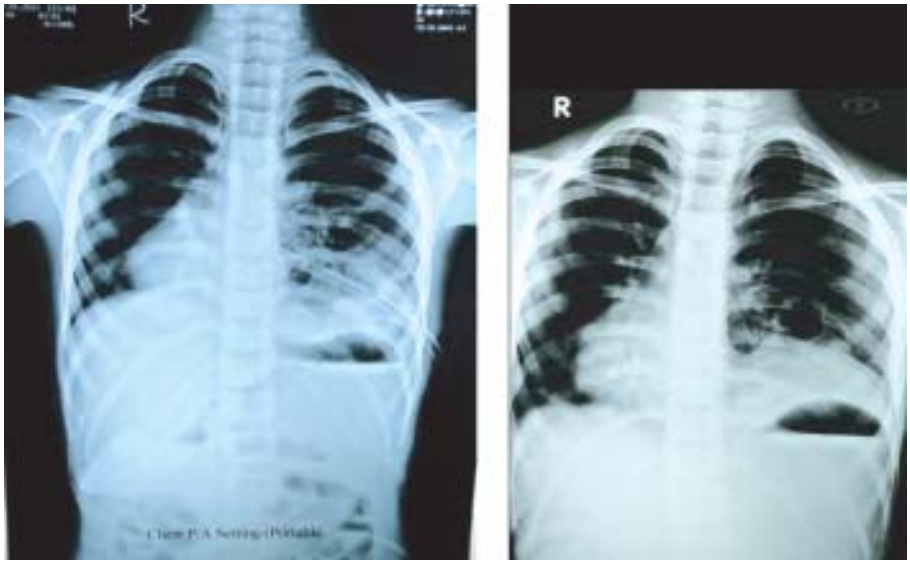

Fig-2: *Post operative day chest radiographs before and after removal of chest drain tube showing extend of lung expansion.*

She was recovering nicely but on fifth post operative day dehiscence of abdominal wound as evident. It was managed by regular dressing and secondary wound closure. She was discharged after three weeks. She underwent closure of colostomy two months later. She had a normal respiratory and gastrointestinal status on next sixth months follow up.

Case Report-2

Two-and a half months old female child presented with history of recurrent respiratory distress and non-bilious vomiting off and on since 2 months. On examination,fullness was present in the epigastrium and left hypochondrium. There was no visible peristalsis. Breath sound was decreased on left lower hemithorax. Laboratory investigations were normal. Chest radiograph revealed multiple air-fluid level into the left hemithorax.

Intraoperatively there was a left posterolateral congenital diaphragmatic hernia with a sac and with organoaxial volvulus of the stomach with the pylorus herniating into the left hemithorax posterior to the fundus.

The contents were reduced, volvulus corrected, sac excised and diaphragmatic defect repaired. Three-point gastropexy was done to anterior abdominal wall. Patient had an uneventful recovery.

Over the last six months she attained a normal growth but experienced three episodes of respiratory tract infections, which improved by medical management. Radiologically there was no evidence of recurrence.

Case Report – 3

A 45 days old male baby presented with the complaints of respiratory distress, and occasional bilious vomiting since birth. The baby was delivered by caesarian section after full term pregnancy. On general physical examination the patient was vitally stable. Abdominal examination revealed mildly distended and tender abdomen in all quadrants with normal bowel sounds. A digital rectal examination revealed normal findings. Laboratory investigations were within normal limits except mild hypokalaemia. Radiographs of abdomen and chest delineated intestinal air shadows in the left hemithorax.

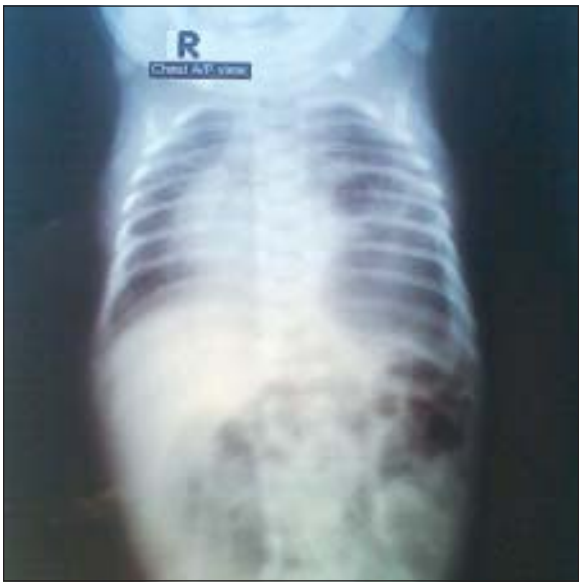

Fig.-3: *Left dome of diaphragm with multiple air-fluid level.*

An exploratory laparotomy was performed by left subcostal incision. The part of stomach, part of small intestine, spleen was found within the left hemithorax herneating through a large defect in posterolateral part of left dome of diaphragm. The left kidney was also found within the thoracic cavity. The stomach, spleen and small intestine were reduced and the kidney was mobilized from the thoracic cavity.

The defect of the diaphragm was repaired with non-absorbable suture material. Patient had an uneventful post operative recovery.

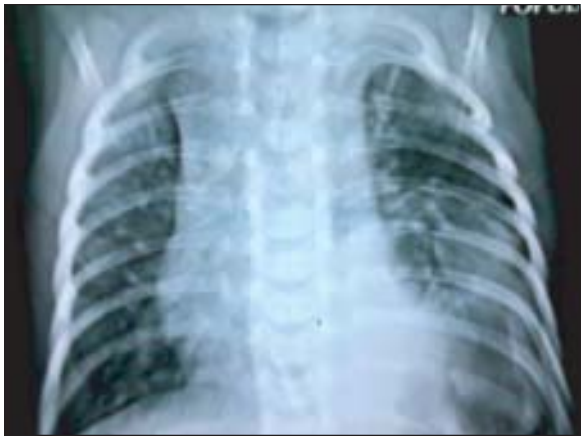

Fig.-4: *Chest radiograph suggestive of diaphragmatic hernia.*

The patient maintained a satisfactory oxygen level and post operative radiograph showed satisfactory lung expansion. The patient was discharged on 8th post operative day. Over the next six months follow up, there was no evidence of recurrence but she developed three episodes of respiratory symptoms,that required medical treatment.

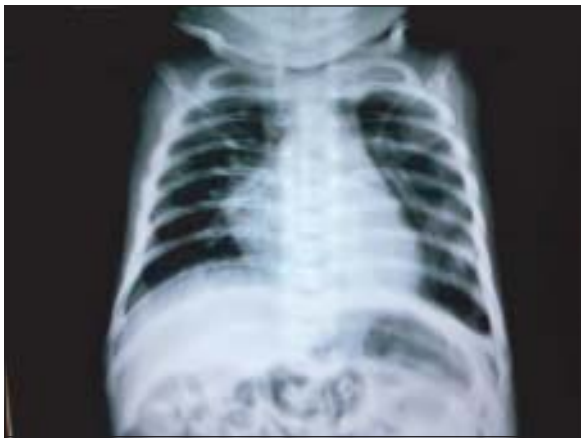

Fig.-5: *Post operative chest radiograph with lung expansion.*

Discussion:

Congenital diaphragmatic hernias mainly present in the neonatal period and are associated with a mortality that has not changed much despite the advances made

in critical care[15]. Rarely, these hernias present later in life, some even in adulthood. There are numerous reports of CDH presenting after infancy. These patients are either asymptomatic or have minimal respiratory symptoms, possibly because the lungs are not hypoplastic. Late-presenting CDH is often difficult to diagnose, and

delays in treatment are common. Moreover, the detection of congenital diaphragmatic hernia may be missed because of intermittent herniation of the abdominal viscera into the thoracic cavity and wide variability in presentation[16]. In addition, Bochdalek hernia may be mistaken for left middle lobe collapse, pneumonic consolidation, pericardial cyst, sequestration of the lung, mediastinal lipoma, or anterior mediastinal mass[17].this might be the reason behind late detection of first case by attending clinician.

Late presenting CDH may present with gastrointestinal tract symptoms that may include intermittent abdominal pain, vomiting, and dysphagia. Respiratory symptoms usually include dyspnea and chest pain. Symptoms may be intermittent or acute depending on the extent of herniation of abdominal viscera into the thorax. An acute presentation is usually due to incarceration, obstruction, or strangulation of the herniated viscera . Diagnosis is ascertained by a combination of chest X-rays, CT and magnetic resonance imaging (MRI), as well as upper gastrointestinal and bowel double-contrast studies[15,18].

A careful analysis of chest films and a thorough search for connecting bowel segments passing through the diaphragmatic defect may help to avoid incorrect diagnosis and an undesirable delay in treatment[19].

We report three cases of delayed presentation of a potentially life-threatening CDH. The variable clinical features of CDH presenting beyond the neonatal period may result in clinical and radiological misdiagnosis. CDH with complicating mediastinal shift and respiratory distress requires urgent gastrointestinal decompression and respiratory support. The most significant factor in achieving diagnostic success is to consider it early in the differential diagnosis to avoid misguided or delayed therapy[20].

Transabdominal and transthoracic approaches have been recommended in Congenital Diaphragmatic hernia repair. The abdominal approach is easily performed through an upper abdominal incision. But when the patient has findings suggesting intestinal strangulation and irreducibiity of the hernia, transthoracic repair of diaphragmatic hernia might be required in addition to laparotomy [21].

Gastric volvulus is rare as stomach is secured in place by the gastrophrenic ligaments, esophageal hiatus, retroperitoneal fixation of the duodenum, short gastric vessels and gastrocolic ligament. It occurs only when these attachments are lax or absent. Secondary causes are eventration ofthe diaphragm, diaphragmatic hernia, congenital bands, wandering spleen and paraesophegeal hernia. Primary volvulus is mainly mesentericoaxial in type while secondary volvulus is mainly organoaxial[21,22]. The clinical symptoms depend on the extent or degree of rotation and gastric outlet obstruction. Operative treatment includes reduction, correction of underlying cause and gastropexy. is not necessary for a successful outcome in secondary volvulus[22]. However, many authors prefer to fix the

stomach even after the correction of underlying defects[15,23].

Following surgical repair of the hernia abnormalities of respiratory function was noticed in most of the cases, which is common as reported by other authors. Significant improvement of lung function is expected over subsequent years of life[12,24].

In most centers Congenital Diaphragmatic Hernia has a mortality rate of 30-60%,[8,12]. Outcomes in present study were more favorable due to absence of other congenital abnormalities. Individual rates vary greatly dependent upon multiple factors; size of hernia, organs involved additional birth defects or genetic problems, status of lung growth, type of treatments, timing of treatments and complications[15,24].

In conclusion, even though diaphragmatic hernia is rare, prenatal USG and post natal chest radiography can diagnose most of cases. Prompt diagnosis and treatment can prevent serious morbidity and mortality associated with complications.

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Ocular Manifestations of a Child with ALL after Chemotherapy

Z RAHMAN[a], J AHMED[b]

(J Banagladesh Coll Phys Surg 2014; 32: 51-52)

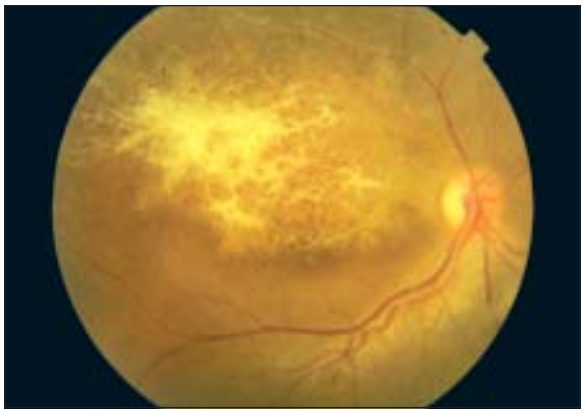

Fig.-1 A: *Fundus photograph of Right eye at presentation shows scared retina in the posterior pole involving macula.*

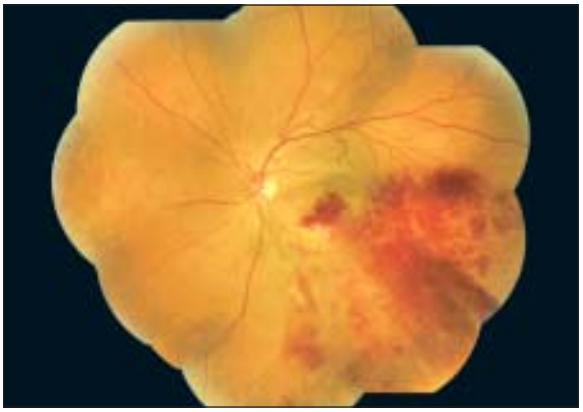

Fig.-1 B: *Fundus photograph of Left eye shows retinal hemorrhage against a background of edematous retina in the posterior pole, also there is vascular sheathing.*

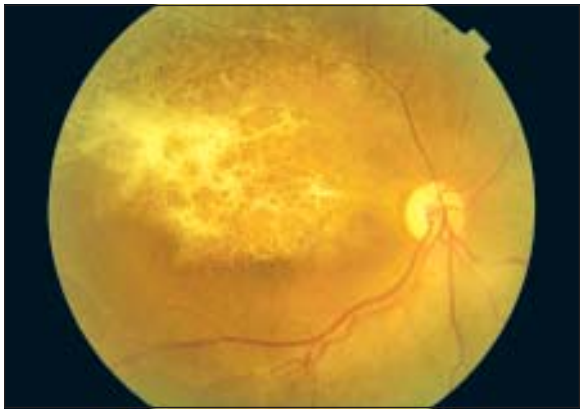

Fig.-2A: *Fundus photograph of Right eye after treatment shows scared thined retina in the posterior pole involving macula.*

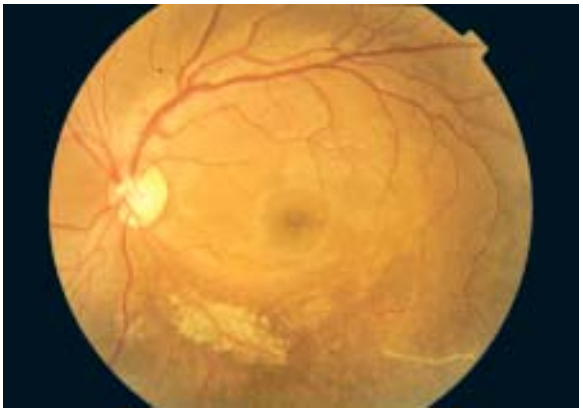

Fig.-2B: *Fundus photograph of Left eye shows healed lesion leaving behind small scar outside the macula.*

1. Dr Md.Zahedur Rahman, Consultant Ophthalmologist and Specialist in Uveal diseases. Bangladesh Eye Hospital Ltd.
2. Prof. Dr Jalal Ahmed, Professor & Head, Department of Cataract, National Institute of Ophthalmology, Sher-e-Banglanagar, Dhaka-1207.

Address of Correspondence: Prof. Dr. Jalal Ahmed, FCPS, Professor & Head, Department of Cataract National Institute of Ophthalmology, Sher-e- Banglanagar, Dhaka-1207.

A nine years old boy presented with sudden loss of vision both eyes. On examination his best corrected visual acuity was HM in right eyes , 6/60 in left eye, anterior chamber was quiet in both eyes, crystalline lens were clear in both eyes, vitreous was clear in both eyes. The fundus examination with indirect ophtalmoscope right eye shows granular area with thinned scared retina in the posterior pole involving macula(Fig:1A), left eye

shows a larger areas of retinal hemorrhage against a background of edematous retina in the posterior pole extending from disc to the vascular arcades, along the distribution of nerve fibers and associated blood vessels (Fig:1B). This type of retinal hemorrhage, retinal edema and vascular sheathing may be due to vasculitis retinae due to any cause of vasculitis (e.g- Eales disease, tubercular, sarcoid, syphilis, SLE, Behcet's disease etc),CMV retinitis. Retinal thinning and scar may be due to any type healed retinal necrosis (Acute retinal necrosis, Cytomegalovirus retinitis, Progressive outer retinal necrosis, Behcet's disease, SLE etc). Clinically we diagnosed as healing CMV retinitis in right eye and active CMV retinitis in his left eye. He is a diagnosed case of acute lymphoblastic leukaemia under chemotherapy since 31 months. He was medicated with vincristine, asparaginasae, methotrexate, daunorubicin, 6-mercaptopurine,cyclophosphamide, Cytosine Arabinoside, oral prednisolone. He was negative for HBs Ag/HIV/HCV. Laboratory tests done by oncologist after diagnosis of CMV retinitis showed blood count within the normal range, erythrocyte hemosedimentation rate of 60 mm, normal C3 and C4, normal urine routine,microcopical examination and absence of proteinuria in the 24 hour collection, positive IgM and IgG CMV serology, positive PCR for CMV from blood. After 4th cycle of chemotherapy he developed ocular symptoms. The induction was initiated with I/V ganciclovir 12 hourly for four weeks followed by I/V ganciclovir once daily for 40 days, up to negative CMV DNA from blood under supervision of oncologist. Subsequently the lesion started to regress. Serial CBC, liver function test and DNA for CMV from blood were done 2 weekly. After 4 weeks of treatment anterior uveitis and vitritis developed which was treated with topical and oral predisolone. Two months after initiation of ganciclovir therapy CMV DNA was negative from blood. The fundus showed large area scarred thinned retina involving macula in right eye(Fig:2A) and small area of scar in left eye sparing macula (2B). The final visual acuity was 6/60 in right eye, 6/9 in left eye.

Discussion:

Cytomegalovirus (CMV) retinitis is a disease which mainly affects patients with acquired immunodeficiency syndrome (AIDS) as well as other immunosuppressed patients, such as the organ transplant recipients under immunosuppressive therapy, those on chemotherapy for malignant diseases, patients with autoimmune disorders like systemic lupus erythematosus (SLE) under immunosuppressive treatment.[1-4] There are a few published series of patient with CMV retinitis without HIV infection.[5,6]

Conclusion:

We believe it is important to inform the existence of this serious and rare clinical complication, especially in our community, where the chronic use of corticosteroids is a routine practice, warning of the irreversible consequences if early diagnosis and treatment are not established.

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Multiple Nodular Swelling in Both Upper and Lower Limbs

A DAS[a], M PARVIN[b], MA KAHHAR[c]

(J Banagladesh Coll Phys Surg 2014; 32: 53-54)

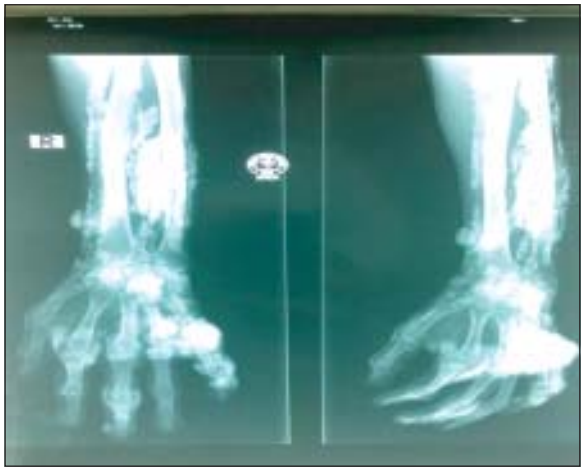

Fig.-1: *Showing Soft tissue calcification involving the forearm and hand*

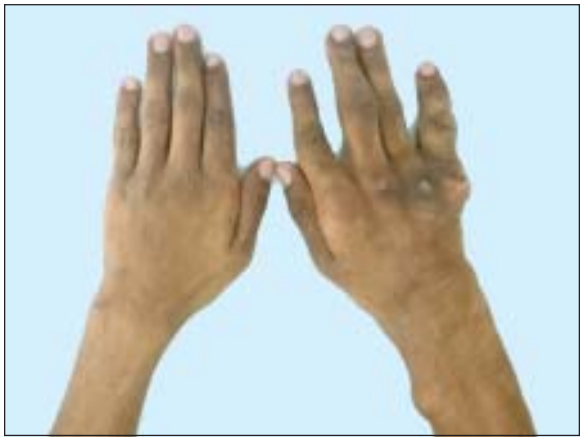

Fig.-2: *Showing nodular swelling with chocky surface in hand and forearm*

1. Dr. Aparna Das. Asso. Prof. Dept of Medicine, Dhaka Medical College
2. Dr. Mania Parvin, Dept of Medicine, Dhaka Medical College
3. Prof. Md. Azizul Kahhar, Professor, Dept of Medicine, Dhaka Medical College

Address of Correspondence: Dr. Aparna Das. Asso. Prof. Dept of Medicine, Dhaka Medical college.M:01914978719

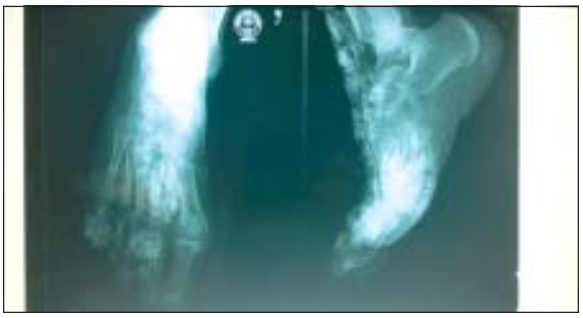

Fig.-3: *Showing Soft tissue calcification involving the foot*

A 17 year- old- boy presented with multiple nodular swelling in right upper limb and feet for 2 years ,and proximal muscle weakness for 2 months. Two years ago patient notice a small nodular swelling over wrist, which was farm later become hard ,with the passge of time he develop multiple nodular swelling without any limitation of daily activities. For last 2 month he develop difficulty in standing from sitting position. Such types of illness didn't run in his family, patient have not histry of taking myopathic drugs . General examination reveals multiple nodular swelling involving ulnar surface of right forearm, 2nd & 5thmetacarpophalangeal joint ,wrist ,right foot , which are nontender , most of them are hard with few firm in consistency, fixed with underlying structure but free from overlying skin without discharging sinus, largest one is (2,2 cm). On CNS examination only muscle power of proximal group of lower limb 4/5 ,feature of proximal myopathy. Investigation shows Hb 11.2g/dl, ESR 15 mm (1stHr), TC 15,000., CPK : 2881 U/L, S. Creatinine, RBS , S.uric acid , S.calcium , S.albumin,Thyroid function test, S. electrolytes and Urine R/M/E are normal. CRP is negative, SGPT : 110 U/L , SGOT : 223 U/L. ANA and Anti-Centromere Ab are negative. Muscle biopsy features consistent with dermatomyositis. Prednisolone 40mg was administered daily with symptomatic improvement. In a recent follow up patient muscle weakness was improved but no exacerbation or resolution of calcinosis was observed.

Discussion:

When calcification is processed any tissue other than bone and teeth is termed calcinosis and can occur in many condition including connective tissue disease, hyper-parathyroidism, renal failure and vitamin D intoxication[1]. Calcinosis may devided into four categories according to the pathogenesis as follows; dystrophic, metastatic, diopathic and iatrogenic. In connective tissue disease, calcinosis is mostly of dystrophic type and it seems to be localized process rather than an imbalance of calcium homeostasis. Calcinosis in connective tissue disease about 9% patient with seleroderma[2,3] and 5% to 20% of adult [4] and 40% to74% of children with dermatomyositis. The existence of calcinosis is indicative of a good prognostic sign of survival but may also be incapacitating.

Dermatomyositis is an idiopathic inflammatory myopathy with characteristic cutaneous manifestation, including helitroperash, gottronpapules, periungual telangiectasias, photo distribution erythema, poikiloderma and alopecia. Although helitrope rash and gottron papules are specific feature, calcinosis may occur upto 40% of children or adolescent.

The laboratory hallmark are elevated creatine kinase, aldolase and transaminase, and a characteristic pattern of EMG-spotty muscle necrosis , regeneration, and inflammation are the pathological hallmark. Calcinosis can be a disability complication that may affect the skin,subcutaneous tissue. It occur most during the course of juvenile dermatomyositis[3]. Calcinosis usually occurred two or three year after onset of dermatomyositis, after that the deposition remain stable and spontaneous resolution has been occasionally reported [5]. The cause and mechanism of calcification are unknown. calcium deposition are ofen in those muscle that were most severly affected during acute phase of disease. Serum calcium, phosphate and urinary calcium values are within the normal range[2] . The calcinosis can be demonstrable both clinically and radiologically. A whole body scan with 99m TC pyrophosphate and CT scan can also identify calcinosis[6]. Aggresive treatment with high doses of prednisolone and physical therapy can decrease the incidence of calcinosis [5] .The use of bisphosphonate in the treatment of soft tissue calcification has varing result[7,8].Two studies shows suppression of GIa synthasis by warfarin sodium may prevent deposition and allow for removal of existing calcinosis. Large and localized mass may be remove surgically[9,10].

Conclusion:

Calcinosis is often signals a improved prognosis. Spontaneous regression of calcification was reported upto 50% of the cases.

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LETTER TO THE EDITOR

(J Banagladesh Coll Phys Surg 2014; 32: 55-56)

To

Editor –in-Chief

Journal of Bangladesh College of Physicians and Surgeons.

Sir,

At first , I would like to thanks to the editor for publishing the original article 'Non HDL Cholesterol versus LDL Cholesterol as a CVD risk factor in Diabetic Subjects' in your journal on October, 2014 issue. I have gone through this article and found the content is very important and informative.The article deals with a facet of the lipid abnormality predominantly seen in our population and hence as cardiologists quite important to us. However, we have to draw your kind attention to some important flaws of the study.

Firstly , the study[1] was evaluating non-HDL cholesterol versus LDL cholesterol as a CVD risk factor in confirmed diabetic subjects. The study design should have been a prospective/cohort study, not a cross sectional observational one.[2] Only then with long term follow up, you can identify those who developed CVD events and compare them with those who didn't. And then taking their different lipid parameters, you can find out the incremental risk of a particular pattern or ratio of lipids regarding CVD risk.

How the authors classified CVD risk individuals in this study[1] were not at all described in this article. Risk calculation is only possible from a case-control study or from a prospective one from OR or RR respectively.[2]

Secondly, even in diabetic individuals, the confounding variable 'sex' has to be taken into account. As Tohidi M et al[3] have shown that in diabetic men, TC, LDL-C, non-HDL-C and TC/HDL-C are independent predictor of incident CVD while in diabetic women, after adjustment for CVD risk factors, only TC/HDL-C ratio resulted in a significant risk for CVD. In the present study, the calculation was not done separately.

Finally, we thank the authors for drawing our attention to the fact that it is not the absolute value of lipids that is important but the proportions of different lipids which are much more important.

1. Prof. Abdul Wadud Chowdhury
Professor of Cardiology,
Dhaka Medical College, Dhaka.

2. Dr. Mohammed Abaye Deen Saleh
Registrar (Cardiology)
Dhaka Medical College Hospital, Dhaka.

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Author Reply

To

Editor-in-Chief

Journal of Bangladesh College of Physicians and Surgeons.

Sir,

We thank Prof. Abdul Wadud Chowdhury and Dr. Mohammad Abaye Deen Saleh for their valuable comments on the original article.[1] We agree that the ideal study design should be prospective/retrospective/ cohort where CVD events can be compared to evaluate the relative contribution of non-high-density lipoprotein (non-HDL) cholesterol and low-density lipoprotein (LDL) cholesterol. But cross-sectional study can also provide some information regarding this.[1] The present study[1] did not compare CVD events, rather two important and established CVD risk factors (LDL cholesterol and non-HDL cholesterol) were compared to include or exclude high risk subjects. In this study[1],

LDL cholesterol up to 100 mg/dL was considered as no/low risk for CVD and LDL cholesterol >100 mg/dL was considered as high risk for CVD, and non-HDL cholesterol up to130 mg/dL was considered as no/low risk for CVD and non-HDL cholesterol >130 mg/dL was considered as high risk for cardiovascular diseases (CVDs). Numbers of subjects at low or high risk classified by non-HDL cholesterol and LDL cholesterol cut-off values described above and mentioned in the article[1] were compared by Fisher's exact test in each group. Sex could have been considered during analysis. But in the background information[2], regarding recommendations of cut-off values of LDL-C and non-HDL-C, sex differentiation was not addressed. Moreover, in methods of this article no category was made based on sex. Here, this cross-sectional observational study[1] aimed to focus on when non-HDL cholesterol or LDL cholesterol is to be the target of CVD risk reduction regarding triglyceride (TG) levels. We concluded that for the detection of high-risk individuals in terms of non-HDL cholesterol above 130 mg/dL and LDL cholesterol 100 mg/dL, LDL cholesterol is a better tool than non-HDL cholesterol at TG concentration up to 150 mg/dL and non-HDL cholesterol is better than LDL cholesterol at TG concentration above 200 mg/dL.

Muhammad Saiedullah
Assistant Professor, Department of Applied Laboratory Sciences
Bangladesh University of Health Sciences (BUHS), Dhaka, Bangladesh
email: md.saiedullah@gmail.com

Shahnaj Begum
Senior Lecturer, Department of Applied Laboratory Sciences
Bangladesh University of Health Sciences (BUHS), Dhaka, Bangladesh

Shoma Hayat
Lecturer, Department of Applied Laboratory Sciences
Bangladesh University of Health Sciences (BUHS), Dhaka, Bangladesh

Syed Muhammad Kamaluddin,
Senior Lecturer, Department of Applied Laboratory Sciences
Bangladesh University of Health Sciences (BUHS), Dhaka, Bangladesh

Dr. Muhammad Rezwanur Rahman
Associate Professor, Dept. of Biochemistry
Delta Medical College, Dhaka, Bangladesh

Dr. Md. Aminul Haque Khan
Professor, Department of Biochemistry
Enam Medical College, Savar, Dhaka, Bangladesh.

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FROM THE DESK OF EDITOR in CHIEF

(J Banagladesh Coll Phys Surg 2014; 32: 57)

Dear Fellows

Let's start this letter with a heartfelt thanks to you for achieving another mile stone. Our favourite journal is now fully online. All Journal activities starting from reading the articles to submission is available in an e-based format. This has opened up enormous opportunity for our fellows, now they can access the journal from any part of the world and will be able to submit and review articles on the go.

We would like our fellows to submit more original articles and request the seniors for quality reviews which will help in improving the quality of the journal further.

Thanks you once again

Professor H.A.M. Nazmul Ahasan
Editor-in-Chief
Journal of Bangladesh College of Physicians
and Surgeons

Obituary

(J Banagladesh Coll Phys Surg 2014; 32: 58)

The following Fellow who died in 2013

Dr. Md. Azizul Islam

Dr. Md. Azizul Islam died on 25th October,2013. He passed fellowship in Medicine in July, 1974 from Bangladesh College of Physicians and Surgeons (BCPS).

Dr. Angel Shubhagata Baidya

Dr. Angel Shubhagata Baidya died on 5th December,2013. He passed fellowship in Surgery in July, 2010 from Bangladesh College of Physicians and Surgeons (BCPS).