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- Editorial Board
The Journal Committee of IJPP wish all our readers a very happy and prosperous New Year. IJPP has completed eleven years since, its inception in 1993. We are grateful to all our reviewers for peer reviewing the articles, all the authors in various field of interest who have contributed articles in our journal and the senior faculty members and experts for replying the queries raised by our readers. We also thank all our readers for their continued support to IJPP.

This issue will focus on “Pediatric Surgery”. The articles were carefully chosen by Dr. V. Sripatii, Co-ordinator for this issue. With his vast experience and rich knowledge in pediatric surgery, he has formulated the topics which will suit to all the clinicians and postgraduates in day to day practice.

In his editorial Dr. Sripatii has stressed that pediatricians and surgeons should work more closely in unison, to provide excellent care to the children of India. Also in his article on dysfunctional voiding in children, he has clarified various disorders and provided a simplified treatment plan. The article written by Dr. Bajpai on Vesico Ureteric Reflux - When is surgery indicated? is a nice review on the current scenario of this condition.

Dr. Srimurthy highlighted the value of pediatric laparoscopy in the management of abdominal surgical problems in neonates and children. In his article on empyema thoracis - recent trends in management, Dr Rasik Shah has discussed in depth the etiopathogenesis of empyema and the role of video assisted thorocoscopic surgery in the management. Early recognition and treatment aspects of respiratory distress in the newborn is well narrated by Dr. Seeniraj.

Various causes of bilious vomiting in newborn is discussed in detail by Dr. Basak and he has also suggested a management algorithm. Dr. S. Venugopal has confined to lower G.I. bleeding in his discussion in the management strategies of bleeding per rectum.

The differential diagnosis in a child presenting with acute painful scrotum is very well reviewed by Dr Meisherii. Dr. Archana Puri et al have described in detail the diagnostic approach to neonatal hyperbilirubinemia with special emphasis on the diagnosis and conditions which require surgical intervention.

The editorial board sincerely thank Dr. V. Sripatii for his valuable contribution and involvement in making this issue on pediatric surgery more interesting to our readers. We welcome our reader’s feedback and suggestion. We also thank all the other authors for contributing their articles in this issue.
CHECK LIST

CHECKLIST FOR INDIAN JOURNAL OF PRACTICAL PEDIATRICS

The checklist must accompany the manuscript.

General

• Original articles which have not been published elsewhere are invited and should be sent to the Editor. They are considered for publication on the understanding that they are contributed to this journal solely.

• Four complete sets of the manuscript are submitted.

• Manuscript is typed double-space throughout with wide margin on one side of paper only including the list of references and tables.

• Manuscript is arranged as follows: Title page, text, acknowledgements, references, tables, figure legends, figures.

• All pages are numbered at the top of the right corner, beginning with the title page.

• The letter of submission has been signed by all authors.

Submission of “Floppy Disc”

Two copies of the articles plus a floppy disc of the wordprocessed manuscript and a list of any nonstandard characters that were used in the disc should be submitted in the usual manner. The preferred storage medium is a 3.5 inch disc in MS-Word compatible format. The publisher is under no obligation to use the submitted floppy disc, but will make every attempt to do so.

Text

• Only generic names should be used

• Measurements must be in metric units with System international (SI) Equivalents given in parentheses.

Title page

• Name and designation of author(s).

• Department where the work was done.

• Running title

References

Identified in the text by Arabic numerals in parentheses.

Type double-space on separate sheets and numbered consecutively as they appear in the text.

Abbreviations of journals conform to the style of Indian Journal of Practical Pediatrics.

Typed as illustrated in “Instructions to authors” with correct punctuation.

Checked carefully by the author(s).

Tables

Numbered with Roman numerals and typed on separate sheets.

Title centered above the table and explanatory notes below the table.

Figures and legends

Not larger than 15cm x 20 cm.

Unmounted and with figure number, first author’s name and top location indicated on the back of each figure. Legends typed double-space on separate sheet. No title on figure.

Each manuscript must be accompanied by a letter of declaration to be signed by each author to confirm that he has seen, read and approved it.

All manuscripts which are rejected will not be returned to author. Those submitting articles should therefore ensure that they retain at least one copy and the reference numbers, if any, of the illustration.

Author’s Signatures.
PEDIATRIC SURGERY FOR THE PRACTISING PEDIATRICIAN

It is with a deep sense of fulfillment that this issue of IJPP on ‘Pediatric Surgery’ is presented. I must thank the Editor-in-Chief Dr. A. Balachandran and my colleagues in the Journal Committee for so readily agreeing to an issue on Pediatric Surgery. It is all the more fitting that this issue will be in your hands during the IAP annual meet in Chennai. In the years to come, I sincerely trust that Pediatric Physicians and Surgeons will work more closely to achieve the aim of providing the very best of care to all the children of India.

The topics have been carefully chosen so that they will be of interest and value to the practicing pediatricians and also will help students in their exams. The authors have been chosen from all over India and are acknowledged experts in their fields. There are many, many more talented pediatric surgeons who could have contributed, but in a journal, space is always a constraint. Depending on the feedback, more issues on topics of your interest in pediatric surgery can certainly be prepared in the years to come.

We trust you will enjoy reading this issue as much as we enjoyed putting it together.

Best wishes for the New Year 2004

Dr. V. Sripathi
and Members of the Journal Committee

NEWS AND NOTES

2nd ASIAN CONGRESS OF PEDIATRIC NUTRITION
January 26-29, 2004

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DYSFUNCTIONAL VOIDING IN CHILDREN

* Sripathi V

Abstract: Dysfunctional voiding represents a group of functional disorders in children, which present with wetting as a symptom. A careful history, voiding diary and basic investigations like renal ultrasound are often enough to categorize the disorder and to prescribe appropriate treatment. In most cases, double voiding, timed voiding, prophylactic antibiotics, anticholinergics and elimination of constipation are enough to eliminate wetting. The most serious variety of dysfunctional voiding is the Hinman syndrome. This results from a persistence of an infantile voiding pattern characterized by voiding against a closed sphincter. This abnormal high pressure voiding creates bladder and upper tract changes in childhood, which are irreversible and indistinguishable from neurogenic bladder disease. This review article aims to clarify the various disorders and provide a simplified treatment plan.

Key Words: Dysfunctional voiding, wetting, children, neurogenic bladder.

Dysfunctional voiding refers to a group of conditions that result in daytime wetting, nighttime wetting and recurrent urinary infection. Before the various disorders are considered, it is important to understand the development of bladder and bowel control.

Natural history of continence

Neonates empty their bladder when a critical stretch threshold is reached by the activation of a sacral spinal reflex. They void about 20 times a day and on an average about once an hour. The pontine micturition center gradually gains control over the act of voiding by the spinobulbospinal pathway and voluntary suppression of micturition becomes possible. This involves two acts, firstly bladder muscle (detrusor) contraction and secondly, external sphincter relaxation. This should happen in a co-ordinated manner for effective emptying.

At 2 years of age, the conscious sensation of bladder fullness is achieved but the need to void is still not under control resulting in physiologic ‘urge incontinence’. From 2 – 4 years of age the bladder comes under voluntary control and by the end of 4 years of age the majority of children have acquired an adult pattern of voiding. The usual sequence of bladder and bowel control is shown in Table 1.

Table 1. Normal sequence of bladder and bowel control

<p>| | |</p>
<table>
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<tr>
<td>Nocturnal bowel control</td>
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<td>Daytime bowel control</td>
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<tr>
<td>Daytime control of voiding</td>
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<tr>
<td>Nighttime control of voiding</td>
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Development of dysfunctional voiding

During the development of continence, if certain ‘bad habits’ are learned they can have a profound effect on bladder and bowel function in later life. Voluntary contraction of the external sphincter is a powerful stimulus to inhibit bladder
muscle. Suppressing the desire to void by voluntarily squeezing the external sphincter leads to dyscoordination between the detrusor muscle and the external sphincter. Effective emptying is not achieved and dysfunctional voiding with its attendant symptoms results. In any discussion of this nature, certain terms must be clearly understood. These have been defined in Table 2.

Table 2. Definition of commonly used terms

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition</th>
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<tr>
<td>Urgency</td>
<td>Need to void immediately with no sensation of bladder fullness.</td>
</tr>
<tr>
<td>Urge incontinence</td>
<td>Involuntary loss of urine associated with strong desire to void.</td>
</tr>
<tr>
<td>Frequency</td>
<td>Voiding more number of times than appropriate for age.</td>
</tr>
<tr>
<td>Posturing</td>
<td>Various positions adopted to prevent loss of urine.</td>
</tr>
<tr>
<td>Infrequent voiding</td>
<td>Less than 3 voids in a day.</td>
</tr>
<tr>
<td>Constipation</td>
<td>Presence of;</td>
</tr>
<tr>
<td></td>
<td>• Fewer than 3 bowel movements per week.</td>
</tr>
<tr>
<td></td>
<td>• Leak of liquid stool in perineum (encopresis) at least once a week.</td>
</tr>
<tr>
<td></td>
<td>• Skidmarks-stool (staining) in underwear daily or several times a week</td>
</tr>
<tr>
<td></td>
<td>• Large wide calibre stools at least once a week that clogs the toilet</td>
</tr>
</tbody>
</table>

Presentation of dysfunctional voiding (Table 3)

Table 3. Presentation of dysfunctional voiding

- Urinary incontinence (daytime or night time)
- Dysuria / Frequency / Urgency
- Urinary infections
- Infrequent voiding
- Incomplete voiding
- Constipation

The main symptom is urinary incontinence without an underlying structural anomaly or an obvious neurological anomaly. Incontinence may be associated with urinary infection with attendant dysuria, frequency and urgency. Holding maneuvers such as leg crossing, squatting or bending from the waist with legs crossed (Vincent’s curtsy) may be employed. Constipation is often an integral part of voiding dysfunction.

History

The following need to be excluded:

- Does wetting date from infancy or has there been a dry interval? If wetting dates from infancy, an anatomical cause such as ectopic ureters needs to be excluded especially in a girl child.
- Have there been any stressful events in the child’s life such as an addition to the family, or unhappy relationships in school?
- Are there any organic causes of wetting such as diabetes insipidus, epilepsy or cerebral palsy?
- Is there any suggestion of sexual abuse?

Voiding/ stooling diary

In order to assess the type of voiding problems and gain a better understanding, it may be useful to maintain a voiding diary. The timing
and volume of each void are recorded, as is the presence of stool in the underpants. The parents should be encouraged to maintain this diary over the weekend. A two or three day record is enough. For e.g. a distinct improvement in dryness over the weekend may suggest that events in school may be the triggering factor.

**Physical examination**

Before concluding that there is no anatomical cause for the wetting, the following need to be excluded:

- The lower back should be examined for signs of occult spinal dysraphism such as a fatty mass, hemangioma, tufts of hair, sinus or abnormal pigmentation.
- A brief neurological examination of the lower extremities and the S2,3,4 dermatomes should be done.
- A normal bulbocavernosus reflex ensures intact lower motor neuron reflex arcs. This is elicited by squeezing the glans penis/clitoris and feeling a contraction of the external anal sphincter.
- Palpation of the abdomen will reveal an enlarged bladder/loaded sigmoid colon.
- Examination of the genitalia will reveal phimosis and meatal stenosis in boys and labial adhesions in girls.
- Redness of the external genitalia in girls may be indicative of vaginal voiding.
- Signs of trauma of the external genitalia in girls may be suggestive of sexual abuse.

**Investigations:**

- Urinalysis and culture will exclude infection.
- Urine calcium/creatinine ratio will exclude hypercalciuria.
- Measurement of urine glucose and specific gravity will exclude diabetes mellitus and insipidus.
- Renal ultrasonography will exclude gross upper and lower tract anomalies and will also indicate if there is a large postvoid residual.
- Plain X-ray of the abdomen will exclude spina bifida (split laminae) and a loaded colon.
- Voiding Cystourethrography can be done if there is suspicion of a neurogenic bladder. This will show vesico-ureteric reflux, bladder trabeculation and will outline the urethra. Postvoid films will confirm if there is an abnormal residual.
- If a suspicion of a spinal lesion remains, MRI of the lumbo-sacral spine is mandatory before completely excluding spina bifida or a tethered cord.
- Urodynamic Studies (UDS) in the diagnosis and treatment of dysfunctional voiding are controversial. In the final analysis, if the type of voiding dysfunction cannot be readily identified or treated, then UDS become necessary. Again, those children who do not readily improve on treatment or relapse need UDS to better characterize the type of bladder dysfunction.

**Voiding disorders and their management**

The following is a brief summary of the various voiding disorders that are encountered in clinical practice with guidelines on management:

**Minor Disorders:**

1. Extraordinary Daytime Urinary Frequency:
   - Seen more commonly in 3–8 year olds.
   - Characterized by sudden onset of urgency
and frequency every 10-20 minutes during waking hours.

# Treatment:

- All caffeine containing drinks and foods should be avoided
- Older children are encouraged to delay voiding by a few minutes when urgency is perceived.

# The condition is self-limiting and usually resolves in a few weeks.

2. Giggle Incontinence (Enuresis Risoria):

# Seen almost exclusively in girls.

# Characterized by complete bladder emptying during giggling or laughing. Caused by a sudden detrusor contraction with sphincter relaxation triggered by giggling.

# Treatment:

- Avoidance of giggling or vigorous laughter in public.
- Anticholinergics, sympathomimetics.
- Methylphenidate

3. Vaginal Voiding:

# Seen in obese girls.

# Due to poor posture during micturition. Urine accumulates in the vagina during voiding and dribbles when standing up.

# Treatment:

- Weight loss
- Voiding with child astride the toilet and face to wall. The back should be straight and legs well abducted.

4. Daytime wetting without other symptoms:

# Usually seen in 3-5 year olds.

# Results from delaying voiding because of intense concentration during playing or while watching TV.

# Treatment:

- Self limiting condition
- Children should be encouraged to void whenever they feel the urge. This retraining establishes a normal pattern in a few weeks.

**Moderate disorders:**

1. Infrequent voiding and Lazy Bladder Syndrome:

# This is seen in girls who void only two or three times a day. The habit is usually acquired by avoiding voiding at school due to fear of the school toilet.

# Wetting is due to overflow, urine stream is poor, voiding is incomplete and infections are frequent. Constipation is usually present

# Ultrasound shows a large capacity bladder with huge postvoid residuals. UDS shows a hypotonic detrusor.

# Treatment:

- Bladder retraining by timed voiding.
- Double / triple voiding to encourage complete bladder emptying and eliminating postvoid residuals.
- Alpha 1- adrenergic blocking agents to relax the bladder outlet and promote emptying.
- Clean intermittent catheterization if the above measures fail.

2. Overactive bladder (Detrusor Instability)
This is the most common form of voiding dysfunction seen in children and has a peak incidence between 5 – 7 years of age. These children have diurnal enuresis with urgency and small frequent voids. They suffer repeated urinary infections and are constipated. Ultrasound shows bladder wall thickening and UDS shows pronounced detrusor contractions during filling. If the pressures exceed 15 cms H₂O, urine leaks out and there is wetting.

**# Treatment:**

- A timed voiding program is instituted every two hours. Prophylactic antibiotics are used to break the cycle of infection and detrusor instability.
- Anticholinergics are given to suppress detrusor instability and allow sufficient time to reach the toilet.
- Constipation should be treated and eliminated before commencing anticholinergics.

**Major disorders:**

1. Hinman Syndrome:

   This represents the extreme end of the spectrum of functional voiding disorders. The condition is known by a variety of other names like nonneurogenic neurogenic bladder, occult neuropathic bladder, psychogenic voiding dysfunction, detrusor sphincter dyssynergia and dysfunctional voiding. The condition is acquired during toilet training when children learn to actively contract the external sphincter. If this is done inappropriately during bladder emptying, dyssynergia develops leading to bladder wall thickening and incomplete emptying (Fig 1). In well-established cases all features of neurogenic bladder disease are present leading to high bladder pressures during voiding and renal damage. If the condition is left untreated, renal insufficiency develops.

   **# Treatment:** This is multimodal and includes:

   - Elimination of constipation
   - Anticholinergics. These help to relax the detrusor and reduce bladder pressure.
   - Clean intermittent catheterization eliminates postvoid residual and prevents infection and also reduces bladder pressure
   - Biofeedback is the modality that directly addresses the issue of bladder sphincter dyscoordination. Children are shown how the sphincter contracts inappropriately during voiding by means of a trace and audio display on external sphincter electromyo-graphy (EMG). They are taught to relax the sphincter during voiding and the effectiveness of therapy is obvious on the EMG.
   - Trans cutaneous nerve stimulation (TENS) is another modality, which is aimed at suppressing inappropriate detrusor contractions. Electrodes are placed on S2,3,4 dermatomes and a low frequency current is passed. This suppresses inappropriate detrusor contractions.
   - In the event of high bladder pressures and progressive upper tract damage augmentation cystoplasty is the only resort.

2. Ochoa Syndrome (Urofacial Syndrome)

   This resembles the Hinman Syndrome with the exception that when smiling is attempted a grimace is produced. This syndrome has an autosomal dominant pattern of inheritance. Treatment is the same as for the Hinman Syndrome.

**Summary** The treatment of dysfunctional voiding involves one or more of the following:

1. Recognition and treatment of constipation.
2. Institution of a frequent / timed voiding schedule.
5. Anticholinergic drugs in the appropriate doses.
7. Clean Intermittent Catheterization to eliminate postvoid residuals
8. In those with established neurogenic damage bladder augmentation surgery.

Bibliography


Fig 1: Hinman Syndrome resulting from detrusor-sphincter dyssynergia. There is simultaneous contraction of external sphincter and detrusor during voiding leading to incomplete bladder emptying, bladder wall thickening, high bladder pressures and upper tract changes.

3. Double / triple voiding to empty the bladder completely.

FIFTH COURSE OF MEDICAL GENETICS & GENETIC COUNSELLING

Date: 9th February to 21st February 2004
Eligibility: Postgraduate in any clinical speciality (limited seats)
Registration Fee: The draft should be in name of ‘Course on Genetic Counseling’ (includes lodging and food Rs. 10,000/- Rs.6000/- for Faculty / Teachers Medical Colleges and Institutes)
For details see: http://www.sgpgi.ac.in/conf/genetics2004.html Write: shubha@sgpgi.ac.in
Send the application and draft to Dr. Shubha R. Phadke, Department of Medical Genetics Sanjay Gandhi Postgraduates Institute of Medical Sciences, Lucknow - 226 014. India.
VESICO - URETERIC REFLUX: WHEN IS SURGERY INDICATED?

* Bajpai M

Keywords: Vesicoureteric reflux, ureteric reimplantation, children

Primary vesicoureteric reflux (VUR) has been reported in one third to one half of patients presenting with urinary tract infection (UTI). The association of VUR, UTI and renal damage is well known. Reflux nephropathy is the cause of end-stage renal failure in 3-25% of children and in 10-15 percent of adults. The indications for surgery in VUR have evolved from the observations made from several studies in the past including some control trials. The general consensus from these studies reveals that 1) antibiotic prophylaxis is an essential part of medical treatment, 2) Unilateral reflux and non-dilated reflux (grades I-III) have a higher chance of spontaneous resolution than bilateral reflux and reflux into dilated ureters, 3) Scars mostly follow breakthrough infections. The treatment of VUR must however be individualized.

The primary goal of the medical management is to provide an infection-free time period using prophylactic antibiotics during which spontaneous reflux resolution can occur without damaging renal function. However questions that remain unanswered include duration of antibiotic prophylaxis, role of asymptomatic bacteriuria and indications for surgery in older children and adolescents.

The ongoing risk of renal damage when patients are being followed on medical management cannot be ignored. Successful surgical correction of reflux decreases the risk of pyelonephritis, eliminates progressive renal scarring and has been associated with resumption of renal growth.

Indications for surgery. (Table 1)

TABLE 1. Indications for surgery

| I] Absolute indications for surgery: |
| 1) Failure of medical management, such as, |
|   i) Breakthrough urinary tract infections |
|   ii) Patient noncompliance |
| 2) A refluxing ureter that opens into a bladder diverticulum |
| 3) Ureteral obstruction in association with reflux |
| 4) Cystoscopic observation of a golf-hole orifice with no submucosal tunnel |

| II] Relative indications for surgery: |
| 1) Presence of massive reflux: Grades IV & V |
| 2) Impaired renal growth or function evidenced by ultrasonography or renal scan |
| 3) In girls whose reflux persists after they have reached puberty, in order to prevent potential problems such as pregnancy-related complications and hypertension. |
Breakthrough infection

Breakthrough infection is development of infection by an organism resistant to the antibiotic used for prophylaxis. Infection rates vary from 10-25% in children in all grades of reflux. All attempts should be made to check the quality of administered antibiotics as this is highly relevant and pertinent in our country while at the same time questioning the compliance of the family. These infections could be entirely asymptomatic or febrile infections. As high as 2/3 of breakthrough infections associated with new scars are reported to be asymptomatic. Two episodes of febrile pyelonephritis or lower tract bacteriuria are considered to be indications for surgery2.

Patient non-compliance

Compliance with nonoperative regime involves maintenance of prophylactic antibiotics, regular urine cultures and repetitive imaging studies to assess the status of reflux, renal function and occurrence of new scars and progression if any of the existing scars.

Compliance rates have been reported to range from 12-90%. While it has been observed that parents of children with high grades of reflux and renal scars are more compliant, the same is not true for lower grades of reflux. The degree of motivation therefore, seems to have a positive impact on the success of this program.

Lower socioeconomic background in terms of economic status, level of education and distance from the treatment centre may be naturally considered as possible contributory factors for noncompliance. However, an analytical review of these factors did not find these important. It may therefore be concluded that patients with breakthrough urinary tract infections, particularly when associated with fever, regardless of the grade of reflux, probably should have ureteral reimplantation.

Grade of reflux and decision for surgery

The decision for surgery varies with the grade of reflux. The incidence of spontaneous resolution while on medical treatment may be as low as 0-9% for grades IV-V of reflux2. In comparison to these, lower grades of reflux, I-III, have been demonstrated to resolve in 5 years in 67%, decrease in 22% and increase in 2%. It can therefore be concluded that, patients with high grade III and grade IV reflux who can be maintained with sterile urine can be followed for a period of time. However, the failure of resolution in those with bilateral reflux is a strong factor in favour of earlier rather than later reimplantation.

Grade V reflux is unlikely to resolve spontaneously. Reflux of this magnitude behaves quite differently from lower grades and is most often referred to as refluxing megaureter. Although, renal scarring is rarely known to occur in refluxing children if a sterile urine is maintained, yet grade V reflux is an exception. This is due to the refluxing megaureter being more analogous physiologically and pathologically to obstructive uropathy.

Available evidence also suggests that primary reflux is unlikely to spontaneously resolve in older adolescents and adults. Therefore, while in lower grades of reflux in infants and younger children every opportunity should be given for the reflux to disappear spontaneously, considerations should change as the age advances.

Higher grades of reflux may often behave differently in the neonate and show resolution. This is considered to be due to the increased compliance of the newborn collecting system. In the first year of life the majority of VUR occurs in boys. Most of these infant boys have increased voiding detrusor pressures and higher grades of reflux than their female counterparts. Through the first several months of life, voiding pressures decrease, detrusor hyperreflexia resolves and
postvoid residuals improve. In these selected patients, the rate of spontaneous resolution is approximately 60% for grade IV reflux and up to 40% for grade V.

**Reflux associated with other anatomical problems such as paraureteral diverticulum**

The size of the diverticulum in relation with the ureteric orifice has an important bearing on the chances of spontaneous resolution of reflux. The presence of the diverticulum interferes with the function of the ureterovesical valve mechanism\(^3\). A smaller diverticulum adjacent but not incorporating the ureteric orifice gives a higher chance for the reflux to resolve. On the contrary a large diverticulum receiving the opening of the ureter does not allow the reflux to resolve spontaneously. Other abnormalities such as cystitis cystica, urethral obstruction and uninhibited bladder may lessen the chance for spontaneous disappearance of VUR. However, they are not contraindications to a trial of medical management.

**Pre-existing scars predispose to higher incidence of abortions and other pregnancy related complications**

Renal scarring, particularly bilateral and renal insufficiency are the real threats to maternal complications and fetal loss\(^4\). Studies suggest that pregnant women with a history of childhood UTIs are at risk for pyelonephritis. Women with hypertension and moderate renal insufficiency are at risk for accelerated decline in renal function as pregnancy advances. Renal scarring, particularly bilateral and renal insufficiency are real threats to fetal loss. However, antireflux surgery does not seem to reduce pregnancy related complications, esp., in women with reflux who had history of childhood UTIs. The incidence of pyelonephritis during pregnancy (17%) in these women is statistically higher than the (4%) incidence before pregnancy. Based upon these reports, it can be concluded that antireflux surgery would be beneficial in adolescent girls with moderate to severe reflux, scarring and renal insufficiency either alone or together.

**PUJ obstruction and VUR**

Whereas the incidence of VUR associated with pelviureteric junction obstruction (PUJ) ranges from 10-14%, the incidence of PUJ obstruction with VUR is reported to range from 0.8-14.0%. For lower grades of VUR medical treatment would be sufficient in most cases. Ureteral reimplantation should be reserved for persistent high grade reflux after successful pyeloplasty.

**Circumcision and VUR**

Infants below 1 year of age have been reported to have a higher incidence of UTI even without VUR. Corroborative evidence suggests that circumcision reduces this incidence\(^2\). Therefore, some reports recommend that circumcision may be helpful in reducing the incidence of UTI in male infants with reflux. This approach may be considered in an individual case only if accompanied by antibiotic prophylaxis. Reports from larger series with long term results are not available to corroborate such recommendations.

**Neuropathic bladder**

Lower grades of reflux have been shown to resolve successfully by prophylactic antibiotics along with anticholinergics to reduce bladder pressures. However, higher grades of reflux have poor resolution rates and these bladders require augmentation cystoplasty to reduce their pressures.

**New scar formation while on prophylaxis**

New scarring occurs due to infection, can occur at any age and with any grade. It may also
occur in adolescents. There is a higher incidence of scar formation below 7 years of age, but in one study 25% children were above 10 years of age when a new scar was seen. However, pre-existing scars can progress at any age and also predispose the kidney to more scar formation. The infection associated with new scar could be febrile, afebrile with lower urinary symptoms or totally asymptomatic.

**Endoscopic subureteric injection treatment:** Endoscopic subureteral injection of tissue-augmenting substances has been studied as an alternative to long term antibiotic prophylaxis and surgical intervention. The biggest advantage of this mode of therapy is ease of use in a Day Surgery setting. Encouraging results have been reported in early grades of reflux. Polytetrafluorethylene was used in the past and subsequently discarded because of migration to distant areas. Newer substances like Deflux are finding wide acceptance after FDA approval in the US. Subureteric injection therapy has the potential of becoming a viable alternative to ureteric reimplantation.

**Future considerations for early surgical intervention**

**The Renin Angiotensin System and VUR**

Children with persistent reflux but preserved renal parameters are managed conservatively with the expectation of spontaneous resolution with age. However, children with persistent reflux of moderate to higher grade (III, IV, V) who apparently have stable GFR, Split Renal Function (SRF) and serum creatinine pose difficulty in management. Although the present criteria for surgical intervention are widely accepted, the long term safety of continued non-operative management has constantly been under scrutiny.

The currently accepted end-points of medical management are inconsistent. Patients who undergo surgery have variable criteria and none of them meet all the criteria for surgical intervention. These criteria are also irreversible. In a recent study we have demonstrated that all children who required surgery as per the internationally accepted criteria, except one (18/19, i.e., 94.7%) had shown an activation and a progressive increase in Plasma Renin Activity (PRA). The mean PRA in this group (n=19) was 6.97 ng/ml/hr versus 3.28 ng/ml/hr in patients who were continued on non-operative management (n=7). Post-operatively, the PRA got reduced and stabilized in all 18 patients at a mean value of 5.4 ng/ml/hr.

Although, in some other congenital uropathies non-operative approach is currently the mainstream yet, there is continued search for early markers for surgical intervention. Study of the Activated Renin Angiotensin System (RAS) is one such marker. Chronic activation of Renin Angiotensin Aldosterone System (RAAS) is associated with adverse structural remodeling of right and left ventricles characterized by reparative (i.e. microscopic scars) and reactive (i.e. perivascular/ interstitial) fibrosis, leading to increased cardiovascular mortality. Even hyperreninemic-normotensive patients carry a 10 to 20% risk of developing hypertension. Identification of early activation of RAS is therefore important since PRA may prove to be of value in early identification of children at risk of developing hypertension and to prevent the deleterious effects of RAAS activation.

Our results suggest that serial measurement of plasma renin activity may help in better stratification of patients with moderate to high grade (III, IV, V) vesicoureteric reflux and reflux nephropathy with respect to management and prognosis.

**Genetic polymorphism and VUR**

Although the detrimental effects of RAS activation are significant, yet there is conflicting
data on the association of Angiotensin Converting Enzyme (ACE) genotype and renal damage. Deletion polymorphism of the ACE gene has been studied as a risk factor for renal damage in patients with congenital uropathies. We investigated the implication of this I/D polymorphism in renal scarring and deterioration of renal function in native Asian Indian children with congenital uropathies. We have demonstrated that the D allele may be one of the genetic susceptibility factors. It may contribute to adverse renal prognosis in patients with congenital uropathies, especially if the anomaly is vesicoureteric reflux. The potential usefulness of genotype evaluation extends beyond prognostication of parenchymal damage to identification of patients who may benefit from angiotensin converting enzyme inhibitors and angiotensin II antagonists.

References


NEWS AND NOTES

MICON 2004 NATIONAL CONFERENCE ON MEDICAL INFORMATICS

January 17-18, 2004

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LAPAROSCOPIC ABDOMINAL SURGERY - UPDATE FOR THE PEDIATRICIAN

* Srimurthy KR

Abstract: Pediatric Laparoscopy is a new and innovative armamentarium available to pediatric surgeons in the practical management of surgical problems in neonates and children. It is based on sound principles of surgery, but with an added advantage of minimal invasion. This article reviews the application of laparoscopy in various abdominal pathologies in children.

Keywords: Pediatric laparoscopy, abdominal surgery.

Laparoscopy (‘laparos-scopie’) means to look into the abdomen through a telescope. Although direct vision laparoscopy was performed nearly hundred years ago for visualization of the abdominal cavity, it was not put to universal use due to poor quality of the telescopes and lighting. Fillip to laparoscopy came in with the availability of Fiber-optic light cable, Hopkins Rod Lens System, and Endo Camera. Philip Mouret in France conducted the first Laparoscopic cholecystectomy in 1987 using the above systems and ever since, the progress in this field has been astronomical.

Pediatric laparoscopy started in the early 90s by a few enthusiastic pediatric surgeons and was mainly confined to diagnostic procedures e.g. Localisation of undescended testis, liver biopsy, appendicectomy, etc. The pediatric surgeons had to do with adult instruments which were long, heavy and unwieldy. In spite of these handicaps, they excelled themselves, overcame the hurdles and began to do a variety of laparoscopic surgical work like Nissen fundoplication, nephrectomy and decortication for empyema thoracis etc. With improvement in the instrumentation dedicated to children, in India a whole range of laparoscopic procedures are being routinely performed.

How does laparoscopy help the Pediatrician?

To take a common example, the following list of conditions in the abdomen may cause diagnostic dilemma.

1. Recurrent abdominal pain (RAP)
2. Evaluation of liver disease.
3. Ascites.
4. Undescended testis.
5. Ambiguous genitalia.

1. Recurrent abdominal pain (RAP)

Children presenting to the pediatrician with RAP is very common. The pain is fleeting in nature and is of short duration. It is mainly located in the umbilicus and seldom associated with constitutional symptoms. RAP in children is often disconcerting to the child and to the family as it can occur at anytime of the day or night and during school hours. In spite of thorough investigations, positive findings are rarely obtained and the pediatrician is in a dilemma as the patient keeps coming back without responding to medication.
Invariably a psychiatric evaluation would have been done with no definitive result. In these circumstances, the child would be referred to a surgeon as chronic appendicitis and the appendix would be removed through conventional open surgery. Through this approach, he would have only limited access to the contents of the iliac fossa. With difficulty, he would be able to palpate the right ovary and uterus. The appendix most often would be normal. Open surgery carries with it, the attendant morbidity of pain, long hospitalization, loss of school days and sports activities. In this situation, diagnostic laparoscopy offers a better yield while trying to locate the pathology accurately with minimal trauma. In addition to the appendix other organs like ovaries, uterus, mesenteric lymph nodes, liver, spleen and peritoneum can be well visualized. The entire procedure is recorded and displayed to the parents so as to assuage their anxieties. Any abnormal pathology can be biopsied. As 5mm ports are used, post operative pain and scar are minimal, hospitalization is short and recovery is fast.

2. Evaluation of liver disease

Recognition of icteric and non icteric liver disease pose major diagnostic problems to child care specialists. Biochemical tests and hepatitis profile studies are valuable indicators of liver pathology but a majority of liver diseases requiring liver biopsy for an accurate diagnosis. Eg: Noncirrhotic portal fibrosis, Cirrhosis of liver of unknown etiology and unexplained hepatomegaly. Conventional liver biopsies are done percutaneously after satisfying coagulation parameters. However due to the blindness of the procedure, the target is often missed. The tissue sample may be inadequate and complications like bleeding and bile leak may be incurred. These problems can be overcome by resorting to diagnostic laparoscopy. Under a short general anesthesia, a 5mm telescope is passed through the umbilicus for locating the pathology in the liver and through another port, a biopsy forceps is passed to obtain a suitable tissue sample and any bleeding spot is coagulated. In addition, the pathologist will have the benefit of the macroscopic appearance by seeing the video recording, which will enable him to interpret the tissue sample.

3. Ascites

Childhood ascites is a vexing problem in clinical practice. When conventional investigations draw a blank, the clinician is in a fix especially if the ascitic fluid is an exudate. Since tuberculosis is a very common disease in our country there is a tendency for these patients to be started on anti tuberculous therapy as a trial. Unfortunately, a number of other diseases mimic this pattern of exudative ascites Eg; omental cysts, mesenteric cysts, ovarian cysts, pancreatic cysts etc. It is essential that a firm diagnosis is established before definitive treatment is commenced. A diagnostic laparoscopy at this juncture will go a long way in clarifying the intra-abdominal pathology. If a cyst is located, the organ of origin is identified, the cyst is de-roofed and the cyst wall is excised for histology.

4. Undescended testis

Impalpable testis is a common problem in clinical practice and a source of anxiety to the parents. Careful clinical evaluation often reveals the location of the testis in the inguinal canal in 80% of children. A small percentage of impalpable testes (13%) can be located with the help of high frequency ultra sound scan. Those in whom the testis is not palpable (20-30%), an assessment has to be done whether the testis is present or not before the age of one year - especially in a child with bilateral un-descended testes. Conventional studies like CT and MRI scans are not helpful in locating intra abdominal testis. Laparoscopy has now become the gold standard for locating these impalpable testes and bringing
them down to the scrotum. In 49% the testis is absent and in 51% it is located in the iliac fossa, pelvis or high up in the retro peritoneum. Laparoscopically, the testis can be mobilized and can be brought down to the scrotum either in one stage or in two stages with very little morbidity unlike open surgery.

5. Intersex disorders

Neonates with ambiguous genitalia, males with severe degree of hypospadias, and females with inguinal hernia require imaging of the internal pelvic organs before correction of these disorders. Although ultrasonography, contrast studies and chromosomal analysis provide information regarding internal anatomy, it is essential that the surgeon must have a complete picture prior to proceeding with surgical correction. A diagnostic laparoscopy done prior to the procedure will give accurate mapping of the internal organs. E.g. In a female child with Testicular Feminization Syndrome, a complete laparoscopic correction can be performed i.e: the absence of uterus can be noted, the intra abdominal testis can be removed and the internal hernial rings can be laparoscopically sutured. Chromosomal males with male external genitalia who have internal female organs can be laparoscoped and the unwanted organ can be removed with minimal trauma.

Laparoscopy, thus has innumerable uses and it is up to the surgeon to rise up in his technical skills and help the referring pediatrician in solving clinical problems with minimal morbidity. It has come to stay and it is high time that the clinicians are aware of the benefits offered by laparoscopic surgery. It offers additional insights to many vexing problems and greatly reduces the morbidity of conventional surgical techniques.

Conclusion

Laparoscopy is indeed a boon in the management of pediatric surgical problems and the benefits are enormous. This new technique has come to stay and it is high time that the clinicians are aware of the benefits offered by laparoscopic surgery. It offers additional insights to many vexing problems and greatly reduces the morbidity of conventional surgical techniques.

Acknowledgements

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NEWS AND NOTES

PRACTICAL PEDIATRIC ONCOLOGY

January 25-26, 2004

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EMPYEMA THORACIS - RECENT TRENDS IN THE MANAGEMENT

* Rasik Shah

Abstract: Empyema thoracis (ET) is a collection of purulent fluid in the pleural cavity. The fact that pulmonary infections were followed by ET and needed external drainage was known to the ancients. Hippocrates, Paul of Aegina, Fabricus and other ancient authorities were concerned with the optimal time and manner of drainage of ET. A high mortality was associated with blood tinged, fetid pus (anaerobic ET). In the 16th century Pare’ manually evacuated a putrid hematoma from the pleural cavity of a French soldier. Kuster in 1889 and Fowler in 1891 performed formal decortication. Thereafter, until the antibiotic era, discussions of therapy largely centered around the relative advantages of open and closed drainage, and its optimal time. In the last decade however, many pediatric surgical centres have started using thoracoscopic drainage/debridement in the management of ET patients. This article discusses in depth the etiopathogenesis of empyema in children and the role of video assisted thoracoscopic surgery in its management.

Keywords: Empyema thoracis, children, video assisted thoracoscopic surgery (VATS)

Natural course of pneumonia and etiopathogenesis of ET

ET most often occurs as a sequel of bacterial pneumonia; however, it can also occur following pulmonary tuberculosis, trauma, intrathoracic esophageal perforation, or operations on the chest. Lower respiratory tract infection in children may get complicated by parapneumonic fluid collections in 50% of patients. These usually resolve with treatment of the underlying pneumonia. Progression to an established ET occurs in 1 of every 155 cases of pneumonia according to one large pediatric series.1

The bacteriology of parapneumonic ET has changed over the years. Staphylococcus aureus was the most common pathogen (92% cases) associated with ET in children below 2 years of age as reported in 1961.2 At present Streptococcus pneumoniae and Haemophilus influenzae Type B are the common causative organisms in bacterial ET. Pseudomonas and other anaerobic organisms have also emerged as important pathogens in sick, hospitalized children and in ET resulting from underlying aspiration pneumonia or lung abscess. ET will remain a clinical problem due to the emergence of drug resistant organisms in the community and persistence of sick, immunosuppressed children from various causes.

Staphylococcus aureus causes a purulent interstitial peribronchial inflammation that results in an abscess formation within the lung parenchyma. These small abscesses coalesce to form a cavity or they can break through the pleura and cause acute pyopneumothorax (ET). An abscess heals with antibiotics leaving pneumatoceles. These have no endothelial lining,
and usually obliterate and disappear without further therapy. Antibiotic therapy will diminish the initial symptoms but allows the disease to progress with a more indolent course until there is a thick pleural exudate. The bacteriology of the pleural fluid has an impact on progression of the ET. ET due to anaerobic organisms have a tendency to form early loculations, which leads to inadequate drainage and often needs surgical intervention. The pleural peel is thicker in streptococcal and Haemophilus ET as compared to staphylococcal ET. The staphylococcus is more likely to form lung abscesses and pneumatoceles without affection of the pleural cavity.

ET is classified into three stages, i.e., exudative, fibrinopurulent and organising stage by the American Thoracic Society. In exudative stage, there is an immediate response with outpouring of thin fluid with a low cellular content, which can be easily aspirated with a needle. The pleura and lung are mobile and the pleural fluid pH, glucose and LDH levels are normal. This stage usually lasts for 24-72 hours. The fibrinopurulent stage is characterised by large quantities of pus posteriorly and laterally, with great numbers of polymorphonuclear leucocytes and fibrin. As the fluid thickens, loculation begins and the lung is progressively less expandable. The pH and glucose values show a fall with increase in LDH levels. This stage usually lasts for 7-10 days. The organising stage is characterised by proliferation of fibroblasts into the exudate on both visceral and parietal pleural surfaces, producing a membranous “peel”. With increasing fibrosis, the ET becomes chronic and the lung is entrapped with inability to expand.

Clinical features

Bacterial pneumonia in children usually presents with cough and fever. Development of pleuritic chest pain and dyspnea heralds evolution of ET. They may also have headache, febrile seizures, and paralytic ileus due to sepsis. On examination breath sounds and respiratory movements decrease on the side of ET. There may be shift of the mediastinum with a dull note on percussion, and a pleural rub. In the late stage of ET, the ipsilateral chest is collapsed with crowding of ribs and obliteration of intercostal space. The mediastinum and diaphragm are pulled on the side of ET along with scoliosis. Rarely empyema necessitans can erode through the chest wall and present with discharging sinus. This is more common with tuberculous ET. Patients can develop pyo-pneumothorax due to air leak either from lung parenchyma (parenchymo-pleural fistula) or by erosion of bronchus (broncho-pleural fistula - BPF).

Diagnosis

Diagnosis is suspected clinically and confirmed by imaging studies. Chest X-ray AP and lateral view and if required decubitus film should be performed to detect the presence of fluid in pleural cavity. Aspiration should be carried out to differentiate between synpneumonic effusion and ET. Presence of air fluid level suggests either anaerobic infection or broncho-pleural or parenchymo-pleural fistula due to either rupture of lung abscess or pneumatocele and very rarely it can be due to esophageal perforation. Multiple air fluid levels suggest presence of loculations. The presence of pneumatoceles suggests infection either due to Staphylococcus or Klebsiella and early loculation suggests infection due to the anaerobic organism.

Aspiration of pus from the pleural space establishes the diagnosis of ET, small or loculated collections may require ultrasound or CT guided aspiration. The fluid is examined by Gram staining, aerobic and anaerobic culture, pH, specific gravity, protein content, glucose estimation and LDH levels. A pH of < 7, glucose
of < 40mg/dl and LDH of > 1000 IU/L is suggestive of ET. These criteria have low sensitivity and cannot be used alone to plan treatment. Gram staining of the aspirated fluid provides information to start appropriate antibiotic therapy, early in the course of the disease. Pleural fluid cultures are often negative either due to previous antibiotic therapy or due to bacterial lysis. Counter current immuno-electrophoresis for antigens to many Gram positive bacteria can help to identify causative organisms in situations where routine cultures are negative. Similarly blood culture and sputum culture should be performed to identify the culprit organism to institute appropriate antibiotic therapy.

Ultrasonography helps to differentiate between pneumonia and fluid collection in an opacified chest. In addition it gives information about loculations and echogenecity of the collection. The CT scan also precisely defines loculations, assesses the underlying lung and degree of pleural thickening. It is also important to remember that the radiological picture may lag behind the clinical stage. Radiographic evidence of pleural thickening may persist for several months even after the complete resolution of ET.

Management

The basic principles in the treatment of ET include control of the offending organism by antibiotics, evacuation of contents and restoration of normal respiratory function by allowing lung to re-expand. The antibiotics are selected empirically and changed according to the Gram staining and culture sensitivity of the aspirated pus. The ET can be evacuated by aspiration, intercostal drain (ICD) insertion with or without fibrinolytics, rib resection, thoracotomy and VATS. After evacuation of the ET chest expansion and compliance steadily improve and breathing exercises hastens the process.

In parapneumonic effusions (pleural fluid pH >7.2, glucose > 40mg/dl, LDH <100 IU/L) appropriate antibiotic therapy and radiological monitoring is required. If the fluid collection is significant causing respiratory embarrassment, aspiration is necessary. Aspiration is done under local anesthesia at the most dependant site or in the fifth intercostal space in the mid-axillary line with a large bore needle (18-20 G), which is placed just above the upper border of the lower rib to avoid injury to the intercostal neurovascular bundle. The aspirated fluid is sent for routine Gram staining, pH, specific gravity, cell count, glucose levels, LDH levels, and culture sensitivity. If fluid analysis confirms it to be empyema and if repeat x-ray chest after aspiration shows incomplete expansion of lung then continuous drainage under water seal is mandatory. In the first few days of exudative stage, it may be possible to aspirate ET completely; however, as the pus becomes thicker and flakes of fibrinopurulent material starts forming, it is not possible to aspirate the ET completely and it will need some other form of drainage.

In the exudative stage and in fibrinopurulent stage, i.e., in the presence of thick pus and flakes of fibrinopurulent material, standard management consists of insertion of ICD along with intravenous antibiotics. A large bore ICD is inserted in the most dependent area and connected to an underwater seal with mild to moderate negative suction (10 to 15 cm water). The milking of the ICD is important to ensure its patency by dislodging small fragments of fibrinopurulent material. The water column should be kept low for good drainage.

Adequate chest drainage and good broad spectrum antibiotics should lead to symptomatic improvement within 48-72 hours. The resolution of fever and leucocytosis with almost complete lung re-expansion as seen on x-ray and/or sonography are favourable indicators during conservative management and chest drainage.
should be continued till drainage decrease to less than 30 ml/day which usually takes 7-14 days. Antibiotic therapy is continued for 2-4 weeks. Successful resolution with conservative management has been reported in 65-82 % of patients, i.e. 18-35 % of them needed thoracotomy with mean hospital stay of 14 to 40 days.\textsuperscript{6-8} If fever persists with inadequate lung expansion then other options should be considered; including irrigation through chest tube, rib resection and pleural toilet, thoracotomy, VATS and instillation of fibrinolytic agents. It is possible to sterilize even loculated collections by use of newer antibiotics; however, bacterial lysis leads to formation of interleukin and other inflammatory mediators, which cause persistent fever and other symptoms. Therefore complete evacuation of ET is vital to ensure the cessation of this inflammatory cascade and symptomatic improvement.

Cyclical irrigation through the ICD ensures patency of tube and may prevent loculations.\textsuperscript{9} To be effective it should be performed in the exudative stage. Rib resection with minithoracotomy can be safely performed in the fibrinopurulent stage of ET even in a very sick patient.\textsuperscript{10} In children the procedure is done under general anaesthesia; about 5 cm of rib is resected at the most dependant area of ET. A finger is inserted in the pleural cavity with the manual breaking of loculations and removal of thick fibrinopurulent material. The procedure is easy and effective during the early stage of ET as the peel is thin and loculations can be lysed with minimal blood loss.

In fibrinopurulent stage with loculation and in organizing stage, ICD or cyclical irrigation are unlikely to work and the child needs either formal decortication or VATS. The term decortication refers to the removal of the thick fibrotic parietal and visceral pleural peel found in the late chronic disease. In fibrinopurulent stage, it is more of pleural debridement and breaking up of loculations which ensures adequate drainage. There is controversy regarding early open surgical drainage; it has benefits of faster recovery and rapid lung expansion. The aggressive approach should be performed in older, immunocompromised children, and in anaerobic infections.\textsuperscript{3} The obvious disadvantages of open surgery are increased morbidity and blood loss.

VATS has changed the outlook in the management of entire spectrum of ET. The term primary and secondary VATS is used for procedures performed before or after a trial of standard management. It is performed under general anaesthesia and selective ventilation of contralateral lung helps in creating a working space. In primary VATS, the first port is placed using an open technique, while in secondary VATS, ICD is removed and the first port is placed through the same opening in chest. The procedure is performed using 5/10 mm and 0/30\textdegree telescopes. Through the first port suction irrigation cannula is placed to suck thin fluid and gentle dissection is performed with it to break loculations; this process creates space to work. Then the telescope is introduced and if needed low pressure (5 cm of water) \(\text{CO}_2\) insufflation at the flow of 1 L/min is instituted to have better visualisation by collapsing the ipsilateral lung. During fibrinopurulent stage, a second small incision is made and open instruments are used to remove thick fibrinopurulent material. The ring (sponge) forceps, stone holding forceps, ovum holding forceps having gentle “S” shaped curve are most useful for rapid removal of the fibrinopurulent material. In the presence of fibrosed thick peel, two or more trocars are placed and the peel is dissected from the underlying lung and removed.

VATS allows determination of the stage of the disease, breaking of all loculi and complete evacuation of thick pus and peel. During early stages it may reduce bacterial load and ensure
that the ICD is inserted in the most dependant position. In addition, VATS gives visual impression of condition of underlying lung, its capacity to expand and presence, site and size of broncho pleural fistula. As thick pus and peel is removed thoroughly, fever resolves quickly (usually in 48-72 hours), ICD is required for short duration (usually for 48-72 hours) and post procedure hospitalisation is reduced to 6-7 days. It also avoids stress related to the insertion of ICD under local anaesthesia. However, VATS needs general anaesthesia, operating room, endoscopic equipment and a team of experienced anaethetist and endoscopic surgeon. Some of the patients undergoing VATS are very sick and they may need careful monitoring in pediatric intensive care units. Primary VATS in ET has been reported to have the best outcome. The author recommends primary VATS if patient presents late (>10 days history) or if there are loculations on imaging studies as standard management is likely to fail. If thick fibrotic peel doesn’t allow dissection to proceed, then open decortication should be performed.

Fibrinolytic agents like urokinase and streptokinase have been used in the conservative management of ET. Allergic reactions to streptokinase are common due to the presence of antibodies in many patients. Incidence of allergic reaction has decreased with the use of newer purified streptokinase. The theoretical advantage of urokinase over streptokinase is that it does not produce antibodies. Patients should be prepared with analgesics and a coagulation profile should be performed prior to the use of fibrinolytics. The diluted fibrinolytics, Streptokinase 12300-136000 U/Kg/dose, can be instilled and the ICD is clamped for 2 hours, and the patient is rotated at 15 minute intervals. The complications of fibrinolytic management is bleeding, BPF and persistence of ET needing some other form of drainage.

**Summary**

To summarise ET is a complex disease, where management needs to be individualized depending upon the presentation and stage of the disease. The author recommends conservative management for a patient who presents early, primary VATS for those who present late or have definite loculations on imaging studies, secondary VATS for patients who have failed to respond to conservative management and open surgery for chronic disease.

**Bibliography**


NEWS AND NOTES

ANNOUNCEMENT

INDIAN ACADEMY OF PEDIATRICS,
Kailas Darahan, Kennedy Bridge, Mumbai - 400 007.

Notice is hereby given that the Annual General Body Meeting of the Indian Academy of Pediatrics is scheduled to be held on Friday, the 9th January 2004 at 17.30 hours at Sri Ramachandra Medical College and RI (DU), Porur, Chennai to consider the following agenda.

Date: October 3, 2003
Place: Mumbai

Dr. Nitin K. Shah
Hony. Secretary General

Agenda:

1. Confirmation of the minutes of the meeting of the Annual General Body Meeting held on 3rd January 2003 at 17.30 hours at Renaissance Mumbai Hotel & Convention Centre, Powai, Mubai.

2. Business arising out of the minutes.


5. Appointment of Auditors and fixing their remuneration.

6. Appointment of Legal Adviser and fixing his remuneration.

7. Any other business notice of which has been circulated with the agenda.

8. Any other business of which 30 days notice has been given to the Secretary General in writing.

9. Any other business with the permission of the chair.
Respiratory distress constitutes the largest cause of mortality and morbidity in the neonatal period. Almost 10-15% of neonates die of respiratory distress. Though, majority of the causes of respiratory distress are medical illnesses, surgical causes amenable to correction may occur in specific groups of neonates. Significant advances have occurred in the management of acute respiratory failure and advancement in the antenatal diagnosis has made it possible to diagnose almost all the conditions inutero. All these advances have contributed to improved results in survival and reduced morbidity.

Recognition of respiratory distress

The following are the signs of respiratory distress:-

1. Tachypnoea - Normal newborns usually breath 40-50 times per minute and rate above 60 is regarded as abnormal

2. Increased work of breathing – If tachypnoea is associated with increased work of breathing, it is called distress. This distress may be mild with subcostal and intercostal retraction or severe with grunting and saw respiration.

3. Central cyanosis – is a late event and occurs when the PO₂ is less than 30-40 mm of Hg.

Various clinical scores have been developed based on the above signs. To evaluate respiratory distress the following aspects should be considered – a) clinical signs b) blood biochemistry and c) chest radiography

Surgical conditions

The conditions which are primarily surgical i.e. amenable to surgical corrections, are as follows:-

Upper air way, nose and nasopharynx: Choanal atresia, nasopharyngeal tumors

Mandibulo facial dysostoses and micrognathia: Pierre Robin sequence

Oral cavity: Macroglossia, cysts and tumours of pharynx

Neck: Congenital goiter, cervical thymic cyst, hemangioma, lymphangioma and inflammatory swellings

Larynx: Congenital microlarynx, congenital laryngeal stenosis, laryngeal web, vocal cord paralysis, cysts and laryngocele, hemangioma of the larynx

Intra thoracic air way obstruction: Tracheal stenosis, tracheomalacia, bronchogenic cysts, tumours

Pulmonary parenchymatous lesions: Congenital lobar emphysema, congenital cystic disease of the lung, lung sequestrations, pulmonary agenesis, hypoplasia
**Pleural conditions**: Pneumothorax, empyema, hemothorax

** Mediastinal conditions**: Pneumomediastinum, mediastinal tumours, duplication cysts, chest wall tumours and malformations

**Anomalies of vessels**: Vascular rings, diaphragmatic hernia, oesophageal atresia

**Abdominal conditions** causing increased intraabdominal pressure

**Principles of management**

1. When a neonate presents with acute respiratory failure, the treatment is urgent regardless of the cause. The following protocol for resuscitation has to be followed:

   - Establish airway
   - Keep the infant warm
   - Provide adequate oxygen
   - Establish vascular line and start IV fluids
   - Assess oxygenation
   - Administer colloids to improve blood pressure
   - Correct acidosis
   - Obtain chest x-ray
   - Continue providing ventilatory support
   - Obtain blood gas analysis frequently every hour until stabilization
   - Assess the neonate and determine the cause

2. When a neonate presents with mild to moderate respiratory distress

   - Investigation particularly CXR is performed to determine the cause.
   - If the condition of the baby deteriorates urgent measures to manage acute respiratory failure are undertaken
   - Surgery is undertaken as soon as possible when a baby presents with mild respiratory distress and the diagnosis is known.

   Ventilatory support and other non-operative measures inherent in the care of neonatal patients like temperature maintenance, prevention of sepsis and correction of coagulopathy should continue in the postoperative period.

**Investigations**

   Chest X-ray is the foremost test done as very few cases can be diagnosed on clinical grounds alone.

   Ultrasonogram, CT chest, barium meal series are done in stable infants when the diagnosis is not clear.

   Though the list of conditions causing respiratory distress is exhaustive, it is beyond the scope of this article to discuss all of them. Only a few conditions have been dealt with in the subsequent paragraphs.

**Choanal atresia**

   This occurs 1 in 5000-10000 births and may be unilateral or bilateral, bony or membranous. In 90% of cases it is bony. In bony atresia the obstruction is located just anterior to the posterior edge of hard palate and in the membranous type it more posterior. The diameter of the choanae and nasopharynx is reduced.

   The common association is maturational dysautonomia and familial incidence is also noted. Respiratory distress is associated with abnormal autonomic control of sucking or swallowing, gastroesophageal reflux and vomiting. They have additional life threatening cardiac rhythm disturbances, hyperthermia, hyperhydrosis and sialorrhoea. Because most neonates are obligatory nose breathers bilateral atresia usually presents as respiratory distress soon after birth. Some of them learn mouth breathing rapidly and
remain asymptomatic but distress is precipitated by attempts at feeding. Unilateral atresia is asymptomatic. The clinical suspicion of bilateral atresia is prompted by inability to pass a catheter through the nose. Relief of respiratory distress is obtained by keeping an oropharyngeal airway and distress gets worsened by pulling the tongue forward and closing the mouth. Confirmation of the diagnosis can be made by instilling radio opaque medium into nostrils and obtaining a lateral skull X ray. CT skull and anterior and posterior rhinoscopy may also be done. The immediate treatment of bilateral choanal atresia is to keep the mouth open by oropharyngeal airway. Transnasal puncture can be done by endoscopy guidance or by blunt dilators followed by stenting and periodic dilatation. In cases associated with syndromes and dysautonomia mortality is high but in isolated cases the prognosis is good.

Pierre Robin sequence

Pierre Robin sequence is a triad of micrognathia, glossoptosis and cleft palate. The hypoplastic mandible is recessed so that the bulk of a normal tongue falls posteriorly and occludes the glottis. With inspiration and sucking the vacuum created in the pharynx exacerbates the airway obstruction. 20% of the patients will have normal palate and another 20% will have associated congenital heart disease. Chronic hypoxia and hypercarbia with acidosis can produce persistent pulmonary hypertension, cor-pulmonale and mental retardation. Patients without cleft palate have highest mortality. Most infants respond to nursing in the prone position, which allows the tongue to fall forward. Passage of a catheter through nasal passage into pharynx negates the vacuum created in the glottic region. Measures to anchor the tongue anteriorly to the lip or mandible are inadequate. If significant obstruction recurs, tracheostomy is needed and gastrostomy is also added to facilitate feeding. With time many infants learn to keep the tongue away from the larynx and in some, significant mandibular growth occurs which keeps the tongue forward. Decannulation of the tracheostomy may be done between 3-18 months.

Laryngomalacia

There is flaccidity of the cartilaginous superstructure of the larynx due to defective deposition of calcium. This flaccid larynx tends to collapse with inspiration and expand with expiration. A number of underlying disorders may be associated with this anomaly. The characteristic symptom is inspiratory stridor with suprasternal and intercostal retraction during inspiration. This stridor may be of variable degree of severity ranging from extreme dyspnoea to normal respiration a short time later. Respiratory infection tends to exaggerate the symptoms. The diagnosis is confirmed by direct laryngoscopy and inspection of the epiglottis and larynx during inspiration. Laryngomalacia usually gets corrected without special treatment and rarely intubation or tracheostomy is needed. Symptoms generally disappear by 1 year of age and avoidance of respiratory infection is essential.

Lobar emphysema

Lobar emphysema or overinflation refers to overexpansion of the air spaces. Depending on the volume occupied by the overexpanded portion, the adjacent lung may be compressed and ventilation compromised. Lobar emphysema may also develop as a complication of barotrauma associated with the treatment of bronchopulmonary dysplasia. Lobar emphysema is considered as a progressive disease process that will produce severe respiratory insufficiency. Symptoms may develop soon after birth but are usually delayed for several weeks or months. Involvement of the left upper lobe is common followed by the right middle lobe and right upper lobe. In 15% of cases there may be associated congenital cardiac problem. Diagnosis is based
on the history, physical examination and chest radiograph. Overdistension of a single lobe is the primary radiological sign. The upper lobe herniates across the anterior mediastinum into the contralateral side. Atelectasis of the adjacent lung parenchyma may be seen. Sometimes the overdistended lobe may be hazy instead of being radiolucent. Treatment is excision of the affected lobe of the lung.

**Congenital cystic adenomatoid malformation (CCAM)**

This anomaly is an uncommon intrathoracic hamartoma resulting from the failure of bronchial buds to join alveolar mesenchyme, characterized by a mass of cysts that may or may not intercommunicate. Cysts are lined by ciliated, cuboidal or columnar pseudostratified epithelium. Cyst wall contains elastic tissue, cartilage, muscles and glandular tissue. Micro and macro cystic forms have been recognized. This anomaly may affect any lobe on one side or may be bilateral. Associated anomalies occur in 20% of cases (Renal agenesis, bowel atresia, diaphragmatic hernia and congenital heart disease).

This condition may be detected in utero by the following diagnostic signs on ultrasonography - 1) compression of the adjacent lung, 2) polyhydramnios, 3) hydrops and 4) placentomegaly. After delivery this anomaly produces symptoms either by compression of the adjacent lung or by infection leading to abscess formation and rarely by spontaneous pneumothorax. Symptoms usually develop in the neonatal period. Evaluation includes chest X-ray and Doppler study. CT, MRI and arteriography are unnecessary. Newborn with distress will need stabilization and measures to reduce persistent pulmonary hypertension. Treatment is resection of the lobe or lobes involved and prognosis depends upon the degree of residual pulmonary hypoplasia.

**Congenital diaphragmatic hernia**

Incidence is 1 in 2500 births. The hernia is on the right in 13%, on the left in 85% and bilateral in 2% of cases. The common form of diaphragmatic hernia is due to failure of closure of one pleuroperitoneal canal (Postero lateral hernia of Bochdalek).

The site and size of the defect, presence or absence of sac, viscera involved and the structure and the margin of the defect, all determine the operative technique, but vital factors which determine the outcome are 1) degree of lung development, 2) effectiveness of pulmonary function and 3) presence of progressive pulmonary disease. Pulmonary hypoplasia is often present on the side of the hernia and there is also variable degree of hypoplasia on the opposite side. Other pulmonary conditions such as atelectasis, pulmonary hemorrhage, infection and emphysema may also be present. In general the clinical picture can be correlated with the state of the contralateral lung. Associated congenital anomalies of the cardiovascular, urogenital, central nervous system and gastro intestinal tract may also occur. There are wide variations in the disturbances of pulmonary function ranging from acute respiratory failure to symptomless dextrocardia and an incidental X-ray diagnosis. The majority of patients with diaphragmatic hernia develop symptoms within first few hours of life. Some are asphyxiated and do not respond to resuscitation. Others have good APGAR scores but soon develop respiratory distress and cyanosis. Occasionally symptoms do not develop for months or years. Cyanosis is usually present and is relieved with oxygen administration. Palpation of the trachea and apex beat indicates mediastinal shift away from the affected side. Breath sounds are diminished or absent but bowel sounds in the chest are usually not audible. The scaphoid abdomen sign is helpful but its importance has been over emphasized. Antenatal diagnosis of
Diaphragmatic hernia is done with increasing frequency using ultrasound. (Polyhydramnios, presence of gastric bubble in the chest). Postnatally X-ray to include whole of chest and much of the abdomen is usually the only investigation needed to confirm the diagnosis. Blood gas analysis and acid base studies are needed during preoperative management. There is no need for urgent surgical correction in diaphragmatic hernia prior to stabilisation. In neonates presenting with respiratory failure, resuscitation is done along the principles discussed in the initial part of this article. In babies who are not responding to conventional ventilatory measures other modes of ventilation like high frequency ventilation or extracorporeal membrane oxygenation may be used if available. If the baby does not get stabilized within 24-48 hours, chance of survival are remote and surgery is pointless. Even in cases which gets stabilized and are taken up for surgery, there will be improvement for 48 hours (Honey moon period) and after that there may be a sudden deterioration which is due to persistent pulmonary hypertension. Ventilation with nitric oxide, surfactant and steroids have been tried but the results are not encouraging.

Overall mortality in cases presenting within 12 hours remains high (50-60%) and has not changed with technological advancements. In cases presenting after 48 hours the prognosis is very good (survival > 80%). Among the survivors long term morbidity is minimal.

Antenatal diagnosis of this condition has made fetal intervention possible. Fetal tracheal plugging through endoscopy has made some progress and its definitive role has not been fully established.

Bibliography


NEWS AND NOTES

BASIC PEDIATRIC INTENSIVE CARE COURSE

January 31st & February 1, 2004

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**BILIOUS VOMITING IN NEW BORN – MANAGEMENT ALGORITHM**

* Basak D  
** Basu AK

**Keywords:** Neonate, bilious vomiting

Bilious vomiting in new born suggests intestinal obstruction unless proved otherwise. However bilious vomiting may be due to sepsis, raised intracranial tension as well as idiopathic causes\(^1\). This article discusses in detail the various causes of bilious vomiting and suggest a management algorithm.

The causes of bilious vomiting are enumerated in Table 1

**Clinical presentation**

Detailed clinical history is essential for differential diagnosis of bilious vomiting. Antenatal history of polyhydramnios is an important clue to duodenal or upper jejunal obstruction. Age of onset of vomiting provides important information regarding etiology. Vomiting on the first or second day of life suggests complete intestinal obstruction either due to intestinal atresia, mid gut volvulus, meconium ileus or total aganglionosis. Vomiting in the second or third week is either due to necrotising enterocolitis, Hirschsprung’s disease (HD) or neonatal sepsis. Associated symptom of inability to pass meconium suggests complete intestinal obstruction whereas delayed passage of meconium is associated with Hirschsprung’s disease. In the newborn, Hirschsprung’s disease may present as acute appendicitis\(^2\). Lethargy, hypothermia or fever are common features of neonatal sepsis. Bilious vomiting associated with passage of blood per rectum usually suggests an acute emergency due to malrotation with mid gut volvulus. Abdominal distension and bleeding per rectum in a premature infant is more likely due to necrotizing enterocolitis.

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**Table 1. Causes of Bilious Vomiting**

<table>
<thead>
<tr>
<th>Category</th>
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<tbody>
<tr>
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<td>Meconium ileus, meconium peritonitis</td>
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<td>Intestinal or mesenteric vein thrombosis</td>
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<td>Anorectal malformations.</td>
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<td>Acquired</td>
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<td>Necrotising enterocolitis</td>
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<td>Lactobezoar</td>
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<td>Brain tumors</td>
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<td>Adrenogenital syndrome</td>
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* Prof. of Pediatric Surgery  
** Consultant Pediatric Surgeon
**Physical Examination**

Assessment of hydration is of utmost importance as vomiting can rapidly lead to dehydration in newborn. Dehydration can be assessed by comparing the baby’s present weight and birth weight and other clinical parameters such as fontanelle, skin turgor, moistness of mucus membrane, urinary output and peripheral circulation. Intestinal obstructions aggravate neonatal jaundice. Visible peristalsis in the epigastrium and absence of distension in the lower abdomen occurs in duodenal or jejunal obstruction whereas lower GI obstruction is associated with generalized abdominal distension. Abdominal tenderness and free gas in the abdomen is associated with peritonitis and intestinal perforation. Incarcerated inguinal hernia in newborn can easily be diagnosed by palpating a tense tender swelling in the inguinal region. Intraabdominal cystic swelling is found in meconium peritonitis with cyst or duplication cyst of the intestine. Examination of perineum is of utmost importance to exclude anorectal malformations.
Fig. 2 Showing double bubble in Duodenal Atresia

Fig. 3 Showing multiple gas and fluid level in ileal Atresia

Fig. 4 Showing calcification in meconium ileus

Fig. 5 Showing pneumoperitoneum
An algorithm presenting a logical clinical approach for evaluating bilious vomiting in newborn (Fig.1)

**Diagnosis**

Several laboratory and clinical investigations are essential for evaluating a newborn with vomiting. Complete blood count, platelet count, C reactive protein, estimation of bilirubin, blood sugar, blood urea nitrogen and estimation of sodium, potassium, chloride and calcium are essential for management. Abdominal roentgenogram in erect posture or cross table lateral view should be taken to detect intestinal obstruction. Distended bowel loops with air and fluid levels are characteristic of intestinal obstruction. Double bubble in abdominal x-ray is diagnostic of duodenal obstruction (Fig.2). Multiple air and fluid levels is suggestive of lower GI obstruction (Fig.3). Calcification is seen in meconium ileus (Fig.4). Peritonitis and pneumoperitoneum (Fig.5) suggest intestinal perforation. Distended loops with soap bubble or ground glass appearance in terminal ileum is suggestive of meconium ileus. Once the obstruction is confirmed, upper GI contrast study is preferred in suspected cases of duodenal obstruction. The lower GI contrast study is of immense value in differentiating microcolon from megacolon, which is a feature of Hirschsprung’s disease. Free passage of dye in the terminal ileum and multiple pellets in the colon suggest meconium ileus (Fig.6). Obstruction to passage of dye in large bowel is due to colonic atresia. Pneumatosis intestinalis and gas in the portal vein are signs of necrotising enterocolitis. Ultrasonography is helpful in evaluating free fluid and blood in the peritoneal cavity. Cystic lesion either due to meconium cyst or duplication cyst causing obstruction can easily be diagnosed by ultrasonography (Fig.7). Antenatal USG is helpful in detecting GI malformation. Meconium plug
in the anorectum is usually relieved by rectal irrigation or during barium enema. Short left colon syndrome, cystic fibrosis and aganglionosis should thereafter be excluded. Anorectal manometry\(^5\) and rectal biopsy will be helpful to diagnose neonatal Hirschprungs disease or neuronal dysplasia.

**Management**

Bilious vomiting in new born should be treated as grave emergency. Prompt attention should be paid to diagnose and to manage the underlying cause. Careful clinical examination and investigations will be helpful in arriving at a diagnosis in majority of cases. Introduction of a nasogastric catheter is of prime importance to decompress the stomach to avoid aspiration. Restoration of adequate hydration is achieved by intravenous fluids and electrolyte abnormalities should be corrected by infusion of calculated doses of electrolytes. Blood should be sent for grouping and cross matching so that it is readily available during surgery. Blood should also be sent for culture if sepsis is suspected. Vitamin K and antibiotics should be started preoperatively and antibiotics should be continued post operatively depending on the culture reports. Laparotomy should be undertaken as early as possible after the baby is resuscitated. Intravenous fluid administration is continued post operatively. Maintenance of calorie and protein supplementation by parenteral alimentation is mandatory as most surgical procedures are associated with prolonged intestinal ileus. Oral fluids and enteral feeding are started when bowel function is restored.

Common conditions causing intestinal obstruction in newborn are described below:

**Intestinal atresia:** Intestinal atresia is the most common cause of intestinal obstruction in newborn. Atresia may be in small or large bowel, small bowel being commoner. Atresia may involve part of the small intestine such as duodenum, jejunum or ileum.

**Duodenal atresia:** Babies with duodenal atresia are usually small for gestational age and have increased incidence of renal, cardiac and anorectal anomalies. Incidence of Down’s syndrome is 30 to 40%. Polyhydramnios is more common in upper GI obstruction and duodenal obstruction can be diagnosed by antenatal ultrasonography\(^4\). Classical double bubble sign in abdominal x-ray is diagnostic of duodenal atresia. Duodenal atresia is treated by duoden-duodenostomy\(^6,7\) or duodeno jejunostomy. Very low birth weight baby may be managed by continuous nasogastric decompression and adequate calorie and protein intake are provided by intravenous alimentation or through feeding jejunostomy. Continuity of the bowel is restored later on.

**Jejunoileal atresia:** Usually have no associated anomalies. The etiology of atresia is thought to be due to intrauterine mesenteric vascular accident. The atresia may be associated with meconium ileus, meconium peritonitis or malrotation of gut. Diagnosis is confirmed by distended loops of intestine with air and fluid levels in abdominal x-ray in erect position. The higher the obstruction the less are the air and fluid levels. Surgical treatment\(^8\) is excision of atretic segment and end to end oblique anastomosis of the bowel. The distended proximal gut is excised to facilitate anastomosis and to regain early peristalsis. In case of hugely distended and foreshortened bowel, tapering jejunostomy is done. In case of atresia with peritonitis resection of the gut and enterostomy is preferred in case of compromised vascularity. Bowel continuity is restored in a second stage surgery. The post operative complications are: anastomatic stenosis, dehiscence of anastomosis and post operative ileus.
**Meconium ileus:** Cystic fibrosis, a common autosomal recessive genetic disorder causes a defect in the exocrine gland secretion. Pancreatic enzyme deficiency has been shown to be the main factor for the development of abnormal meconium. Viscid tenacious meconium obstructs the terminal ileum. The distal ileum and colon contain inspissated pellets of meconium. The entire large gut is of small caliber due lack of use. Abdominal radiograph shows distension of small gut with no air and fluid level. Barium enema shows microcolon and dye is obstructed in the terminal ileum. Antenatal perforation of the distended gut produces meconium peritonitis and calcification. Accumulation of meconium with fluid produces a meconium cyst. Simple meconium ileus can be managed by therapeutic hyperosmolar enema with gastrografin. The hyperosmolar enema draws fluid into the intestinal lumen and dislodges viscid meconium and helps in the expulsion of meconium. Adequate intravenous fluid should be infused to combat fluid loss and dehydration during this procedure. The complications of gastrografin enema are shock, intestinal perforation, necrotising enterocolitis and death. Meconium ileus which does not respond to therapeutic enema is managed by laparotomy. Enterotomy and irrigation of the bowel is done to remove the viscid meconium. The dilated ileum is excised and gut is exteriorized or various types of anastomosis are done to restore bowel continuity. Post operatively N-Acetyl-cysteine is administered through nasogastric tube to facilitate expulsion of meconium.

**Hirschsprung’s disease**

Hirschsprung’s disease or congenital aganglionosis occurs in 1 in 5000 to 7000 live births. This condition is caused by failure of migration of ganglion cell from neural crest to the colon. The failure of migration of ganglion cells affects varying length of colon, the most common site being recto-sigmoid region. As a result the anal sphincter becomes spastic and proximal colon dilates to overcome the obstruction. The classical symptoms are constipation and abdominal distension and newborn presents with vomiting and intestinal obstruction. The classical features of narrow distal colon, cone and dilated proximal segment in barium enema is rarely discernible in new born. Anal manometry, a diagnostic procedure which shows absence of relaxation of anal sphincter in response to distension of rectum, is operator dependent and has limitations in new born and premature infants. Rectal biopsy is the gold standard for the diagnosis of Hirschsprung’s disease. Absence of ganglion cell in the submucus and myenteric plexus is the hallmark of diagnosis. Rectal suction biopsy with acetyl cholinesterase

**Malrotation of intestine**

Abnormalities of intestinal rotation represent a spectrum of diseases caused by failure of normal intestinal rotation in different stages in embryo. Intestinal obstruction due to malrotation is caused by volvulus of gut or bands. The circulation to the gut is jeopardised in volvulus leading to gangrene and presents with bilious vomiting, abdominal distension and profuse rectal bleeding leading to shock. It is termed as volvulus neonatorum in new born. Diagnosis is confirmed by straight x-ray abdomen and rarely upper GI contrast study is necessary. In volvulus neonatorum immediate surgical treatment is required after resuscitation of the baby. The volvulus is reduced by derotating the gut, gangrenous bowel and bands are resected and end-to-end anastomosis is done to restore the continuity of the bowel if the gut is viable. In case of doubtful viability of the gut enterostomy is done and bowel continuity is restored in second stage. In case of viable gut, the Ladd’s bands are resected to decompress the duodenum and other adhesions are released and caecum and large bowel are placed on the left side and small bowel on the right side.
histochemistry improves the diagnosis in newborn\textsuperscript{12}. The primary aim of treatment of Hirschsprung’s disease in new born is to relieve obstruction. If the obstruction is not relieved by rectal irrigation by normal saline laparotomy should be done and colostomy should be performed above the level of obstruction. Once the obstruction is relieved either by colostomy or by irrigation the basic principle of surgical treatment is excision of aganglionic segment and anastomosis of the ganglionic bowel to the rectum. There are several procedures to achieve this goal. The procedure is to be undertaken when the bowel is completely decompressed and baby is fit for major surgery.

**Anorectal malformations:** Anorectal malformations occur approximately in 1 in 5000 live birth. The incidence is higher in male infants and clinical presentation are variable from complete obstruction to fistula formation with the

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**Fig. 8 Invertogram showing intermediate anomaly in anorectal information.**

**Fig. 9 USG showing hypoechoic lesion in duplication cyst of intestine.**
bladder, urethra, vagina and perineum. The common associated malformations are esophageal, cardiac, renal and skeletal anomalies. The lesion can be diagnosed by perineal examination and abdominal x-ray. The level of obstruction can be diagnosed by invertogram (Fig 8). USG, CT and MRI. In case of high anomalies initial treatment is colostomy and perineal anoplasty is performed in low anomaly. In majority of females, rectum opens into vagina and reconstruction is done later by different operative procedure.

Necrotising Enterocolitis (NEC): NEC results from intestinal mucosal injury due to low blood flow status and hypoxia with secondary bacterial invasion. Prenatal and intrapartum complications usually lead to hypoxia in fetus. Prematurity and LBW are predisposing factors. The bowel is markedly distended with areas of inflammation and necrosis with mucosal ulceration are seen. The terminal ileum is mostly affected and necrosis may lead to gangrene and perforation. The treatment protocol includes correction of hypovolemia, hypoxia and infection. Surgery is indicated if complications supervene such as perforation, gangrene of intestine and abscess formation. Excision of non-viable gut with enterostomy is the operation of choice, but in very poor risk patients peritoneal drainage may be attempted.

References


BLEEDING PER RECTUM IN INFANTS AND CHILDREN- MANAGEMENT STRATEGIES

* Venugopal S

Keywords: Bleeding per rectum, children

Bleeding from the rectum, a distressing problem for the parents, presents a major challenge to the Pediatrician and Pediatric Surgeon in terms of diagnosis and management. The etiology of gastrointestinal (GI) bleeding in children is different from that of the adults. The amount and rate of the blood loss will determine the urgency with which the problem has to be tackled. One should also bear in mind that in about 10% of children, the cause will be located in the upper GI tract. This discussion will be confined to the causes and management of lower GI bleeding.

The age of the child, quantity, rate and quality of the blood loss and associated symptomatology needs to be taken into account when trying to arrive at a diagnosis and plan management.

Age: In the neonate and infant, congenital causes such as malrotation with volvulus, Meckel’s diverticulum, tubular duplications and vascular malformations are likely important causes. In a stressed, especially premature infant, splanchnic hypoperfusion due to ‘Dive reflex’ will raise the risk of Necrotising Entero-Colitis (NEC).

Haemorrhagic disease of the newborn is not an infrequent problem in the neonate, especially when Vit. K is not routinely administered. Simple local causes such as anal fissures are fairly common. In neonates, one also needs to consider the possibility of swallowed maternal blood, either at the time of delivery or from a cracked nipple in the case of breast fed infants. As age advances, acquired causes, with or without underlying congenital factors become more common. Anal fissures continue to remain a problem. Intussusception, Juvenile polyps, bacterial, viral and helminthic infections, inflammatory bowel diseases, vasculitis and coagulopathies appear in the list (Table 1).

Quality: Fresh red blood indicates a very distal source of bleeding while altered blood indicates that the blood has stayed in the GI tract for a period before expulsion; the longer the time taken the greater being the alteration. The rate of transit of blood will also affect the degree of alteration. With brisk haemorrhage and rapid transit, even a bleeding duodenal ulcer can present with rather fresh blood per rectum. Presence of mucus along with blood indicates colonic inflammation. Loose stools with mucus and blood would suggest enteritis with or without colitis. Blood and mucus alone without stools should indicate not only inflammation but also obstruction as in intussusception.

Quantity: Small streaks of blood smeared on the outside of stools indicate a low, localised source of bleeding, as in anal fissure. Drops of
blood at the end of defecation also indicate a rectal or anal lesion such as polyps or hemorrhoids. Blood mixed with stools suggest a more proximal location of the pathology. Large bleeds are usually seen in Meckel’s diverticulum, duplications or vascular malformations. Beware of small spotting, as this may be a forewarning of an impending large bleed.

Associated symptomatology: Pain is a very useful clue to indicate the source. Pain on defecation indicates a lesion in the anal canal. Associated abdominal pain would indicate inflammation or obstruction of the bowel. Colicky abdominal pain usually indicates obstruction and associated lower GI bleeding should be viewed with concern, as this indicates a possible vascular compromise; volvulus and intussusception being the most important considerations. Painless bleeding is suggestive of Meckel’s, duplication or vascular malformations.

Appearance of a mass per rectum can be a polyp or rectal prolapse. Rarely a prolapsing intussusceptum can mimic a rectal prolapse. Simple digital examination to verify that the anal verge has everted out (inability to insert a finger between the mass and the anal canal) will confirm prolapse of the rectum. Systemic features of infection may indicate an infective process. Predisposing factors leading to intestinal hypoxia such as stress, respiratory distress syndrome, cyanotic congenital heart diseases, early large volume feeds and exchange transfusion, especially in premature infants, points to possible NEC. Chronic constipation from birth points to Hirschsprung’s disease and associated enterocolitis. Enterocolitis can also be antibiotic induced. Evidence of vasculitis or bleeding from other sites would suggest generalised disorders as the underlying cause such as Haemolytic Uraemic Syndrome or Henoch Schonlein Purpura. But it must be borne in mind that GI

<table>
<thead>
<tr>
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bleeding may be the sole manifestation of a coagulopathy. Peripheral vascular malformations may be a clue to similar pathology elsewhere. Congenital abnormalities associated with syndromic pathology such as peri-oral, digital pigmentation and exostosis may suggest intestinal polyposis.

**Management:** Evaluation of the clinical status of the child with special reference to cardiovascular stability and estimation of the rate and extent of blood loss takes priority. In most cases, the blood loss is slow and intermittent, allowing time for elective evaluation and management. In major bleeds, IV access should be established and volume replenishment and stabilisation achieved before detailed assessment and investigation.

**History:** Apart from enquiring about symptoms referred to earlier, history regarding drugs taken and food ingested by the child and, in the case of breast-fed infants, by the mother should be obtained. History of trauma, locally as well as to the abdomen will be helpful. Family history of bleeding diathesis, polyps and inflammatory disorders should be looked into.

**Examination:** General examination starts with looking for evidence of acute or chronic blood loss in the form of hypovolemia or anemia.

The abdomen is examined for signs of obstruction, inflammation or mass (intussusception). Visual and digital examination of the anal canal and rectum, supplemented by a proctoscopic examination will reveal local pathology such as anal fissures, prolapse and polyps (60% within reach of finger). An auroscope or nasal speculum is suitable for proctoscopy in infants. Physical examination is completed by looking for indirect clues as well as associated pathology, discussed above. If upper GI bleed is suspected, passing a naso-gastric tube to check for presence of blood in the stomach is a simple and useful step. Presence of blood in the stomach will direct the search towards causes in the upper GI tract.

**Investigations:** Except in cases where the cause of the bleeding is evident, investigations are essential. If in any doubt, the red or black colour should be confirmed to be due to blood and that, in the case of infants, it is not ingested maternal blood.

Complete hematological evaluation, including coagulation profile, is necessary. Stools can be examined for evidence of infections, infestations and inflammation.

**Imaging:** Ultra sound examination of the abdomen and pelvis is the most valuable and least invasive imaging technique, especially when looking for mass lesions as in intussusception. Doppler may further add to its usefulness in evaluation of vascular malformations.

**X-ray** of the abdomen will help to identify features of obstruction or inflammation. When duplication anomalies are suspected, x-ray of the spine may provide an indirect clue in the form of vertebral anomalies.

**Contrast enema** for evaluation of lower GI bleeding must be performed as a double contrast study with adequate preparation, except in the case of suspected intussusception where this is not only diagnostic but also therapeutic.

When malrotation is suspected, an upper GI contrast study to assess the duodenal C-loop is preferred over a barium enema to locate the caecum, since the caecum in infants can be very mobile.

**Radioisotope imaging** with Technetium pertechnate is useful in detecting heterotopic gastric tissue in Meckel’s diverticulum and duplications, with an 85% success rate. ‘Bleeding scan’ using radioisotope tagged RBCs can help
to locate the site of bleeding if the rate of blood loss is 0.1 ml/minute or greater.

**Angiography** is useful when vascular malformations are suspected and to locate the site of bleeding. However the rate of bleeding has to be more than 1ml/minute for successful localisation and hence less sensitive than the ‘bleeding scan’.

**CT scan** is indicated in mass lesions and also in evaluation of trauma.

**MRI**, although non-invasive and useful, the requirement of anaesthesia or sedation, makes it a less attractive tool in small children.

**Endoscopy** has the advantage that it not only visualises the pathology but also allows therapy or biopsy. Flexible sigmoidoscopes and colonoscopes have made endoscopy easier and allows more complete evaluation. In an emergency, the unprepared bowel and presence of blood will impair visualization.

Upper GI endoscopy will be needed when an upper GI cause needs to be excluded. Futuristic ‘Capsule video-endoscopy’ of the GI tract provides an opportunity to visualise otherwise inaccessible areas of GI tract and may become a valuable addition to investigatory tools in children old enough to swallow the capsule.

**Laparotomy / Laparoscopy** can be considered as the final diagnostic step when all other investigations fail to identify the cause. External examination of bowel can be supplemented by simultaneous internal visualisation by intra-operative colonoscopy in difficult cases.

**Conditions causing lower GI hemorrhage**

**Anal fissure:** Passage of hard stools is the usual etiological factor for this common cause of bleeding per rectum in infants and children. Associated pain makes the child withhold stools longer making the stools harder, thus setting up a vicious cycle. Chronicity comes from repeated trauma. Diagnosis is evident from blood streaks on the surface of the stool. The fissure and its tell tale sign of sentinel skin tag will give away the diagnosis without the need for additional investigations. Keeping the stool soft with diet and laxatives, sitz bath and topical pain-relieving ointments will help to heal the fissure. Only when these fail does one need to resort to surgical interventions such as excision of the fissure and sphincterotomy or anal stretch.

**Polyp:** Juvenile polyps are the most common GI tract polyps in children. They are single or only a few and mostly distributed in the rectum and distal colon. The natural history consists of heaping up of epithelium, which develops into granulation like tissue, acquires a stalk and eventually gets auto-amputated. Therefore juvenile polyps manifest between 5yrs and 15 yrs of age. These self-limited polyps have very low malignant potential. Simple polypectomy is curative. Polyps associated with Peutz-Jeghers syndrome are hamartomatous polyps. Although malignant potential is low, risk of recurrent intussusception is high. 14% of girls can develop ovarian tumours. Syndromes such as Gardner’s and Turcot’s have adenomatous polyps with potential for malignant transformation. Familial polyposis coli have adenomatous polyps with a high propensity for malignant transformation. Hence all the polyp bearing epithlium must be eliminated by total procto-colectomy. Juvenile polyposis coli has numerous polyps affecting the entire GI tract, but dominantly colon. It presents with chronic massive bleed and protein loss that adversely affects the child’s growth and development and is treated by colectomy. Lymphonodular hyperplasia, appearing as nodularity on endoscopy and umblicated lesions on double contrast studies, are seen between the ages of 1 year and 5 years (peak between 3rd and 4th year). These are self-limited benign lesions and need only supportive therapy.
Prolapse of rectum

May be idiopathic or secondary to helminthiasis (Trichuris) or underlying neuromuscular deficiency (myelomeningocele). Both diarrhea and constipation contribute to its development. Sitting on toilet seats with large holes is another factor. Removal of underlying etiological factors, nutritional resuscitation along with keeping the prolapse reduced will result in resolution. Only when these measures fail, should one consider surgical intervention such as Thiersch suture, submucosal sclerosant injections and rectopexy to sacrum.

Meckel’s diverticulum and duplications: Only those Meckel’s diverticulae with heterotopic gastric tissue can cause peptic ulceration that results in GI bleed. Tubular duplications which have similar heterotopic tissue can also cause peptic ulceration and behave like a Meckel’s diverticulum. Typically they present with painless passage of large amount of chocolate colored blood (slightly altered). Surgical removal of the diverticulum or duplication along with the ulcer bearing area near the mouth of the lesion is essential to arrest the bleed.

Volvulus with ischemia: Midgut volvulus is the most common type of volvulus in infancy. Malrotation, which results in narrowing of the base of the mesentery due to the proximity of duodeno-jejunal flexure and ileo-caecal valve, is the usual cause. Acute or intermittent obstruction during infancy, especially associated with severe abdominal pain should alert one to its possibility. Bleeding per rectum in such a clinical setting is an ominous sign of impending ischemia and expeditious surgical management is the only life saving step. At laparotomy, undoing the volvulus and Ladd’s procedure to correct the malrotation is done. In extensive ischemia, after correction of volvulus, temporary closure of abdomen with a planned ‘second look’ after a 24 to 36 hour period of intensive resuscitation and support, may permit salvaging of some of the bowel with borderline ischaemia.

Rarely volvulus of midgut can occur even with normal rotation. Volvulus of isolated ileal loop along the axis of a vitello-intestinal duct or band and caecal volvulus are other rare types of volvulus encountered in children.

Intussusception: Idiopathic intussusception is usually seen between the ages of 5 months and 24 months. Outside this age group, secondary intussusceptions due to an underlying lead point should be suspected. Colicky pain, vomiting and passage of blood and mucus (‘redcurrant jelly’) without stool are the classical clinical triad. Since it may take time for all the signs to appear, a high index of suspicion should be maintained to make an early diagnosis. With early diagnosis, hydrostatic reduction, using air or saline or barium under ultra-sound or fluoroscopic monitoring, has an 80% to 90% chance of success. Surgical exploration is required only when hydrostatic reduction fails or a lead point is suspected. At surgery resection is required only for irreducible or ischaemic intussusceptions and also for removal of a lead point. While in infancy congenital lesions like Meckel’s or duplications are usual lead points, in older children acquired lesions such as polyps and GI lymphoma are to be considered.

GI infections: Salmonella, shigella and campylobacter enteritis can present with abdominal pain and loose stools. Blood and stool cultures and serological tests will establish diagnosis and antibiotic therapy will be curative. Supportive therapy will be needed until resolution is achieved. Need for surgical intervention to stop the bleeding is exceedingly rare. Yersinia pseudotuberculosis and TB are other bacterial infections that need to be kept in mind. Rotavirus is a common viral agent that causes gastroenteritis with bloody stools.
In broad spectrum antibiotic therapy, pseudo membranous enterocolitis due to Clostridium difficile is a distinct risk. Proctoscopic demonstration of a pseudo membrane will help in diagnosis and Vancomycin and Metronidazole are effective therapy. Entamoeba coli, giardia and trichuris trichura are common causes of colitis. With effective antihelminthic drugs currently available, elimination by drug therapy is easy and effective.

GI inflammation: Although rare, inflammatory bowel disease can occur in infancy and childhood. Management with Salazopyrin along with short supplements of steroids to cover acute exacerbations, is effective. Surveillance of the colon with colonoscopy and biopsies is essential to detect dysplasia which will indicate the need for surgical intervention.

Necrotising Enterocolitis (NEC) in Neonates is probably the result of mucosal damage from multifactorial insults. Enterocolitis can also complicate underlying conditions such as Hirschsprung’s disease. Enteric mucosa could be involved as part of generalised vasculitis as in Henoch-Schonlein purpura or hemolytic uremic syndrome.

Allergy to milk feeds, can cause GI bleeding, but must remain as a diagnosis by process of exclusion of all other causes.

The management will be removal of the offending agent and treatment of the underlying inflammation.

Vascular malformations: The vascular malformations located in the colon or small bowel are likely to present with lower GI bleed. Co-existing peripheral haemangiomatous lesions (in 50%) may be an available clue. Endoscopic therapy (sclerosants, thermal or laser photocagulation) or embolisation may be applicable only in a select few, even when the expertise and equipment are available. More often surgical resection of the affected segment will be needed. Hemorrhoid as a cause of rectal bleeding is extremely uncommon in children. When seen, congenital vascular malformations and underlying portal hypertension with porto-systemic shunting should be kept in mind, as possible causes.

Coagulation disorders

Coagulopathies can manifest with GI hemorrhage as the sole presenting feature. A comprehensive evaluation of coagulation is mandatory in completing the work-up of a child with GI hemorrhage where no cause is evident. Correction of underlying defect will arrest the bleeding. When detailed evaluation of the coagulation cascade is not available, fresh blood transfusion along with Vit.K will be an imprecise but effective way of stopping the bleed.

Trauma: Local trauma to rectum and anal canal as well as trauma to abdomen and acceleration or deceleration injuries (as in fall from a height) can cause bowel damage. For extensive damage or associated perforation surgical intervention will be required. Blunt trauma to liver can cause haemobilia which will manifest as lower GI bleed. CT scan and upper GI endoscopy will help in arriving at the diagnosis. In persistent haemobilia embolisation or segmental resection of the damaged segment may be needed.

Summary

Lower GI bleeding can have a wide spectrum of underlying aetiology as well as in the amount and rate of blood lost. We should also bear in mind that causes of upper GI bleed can also present with lower GI bleed. It can also be the manifestation of systemic disease. Hence a detailed history and a complete physical and hematological evaluation are essential. Fortunately most of the rectal bleeding in children are small
bleeds from detectable benign, local lesions. Association of acute emergencies like malrotation with volvulus and intussusceptions mandates the need for a high index of suspicion and immediate management. But at the other end of the spectrum, in a group of children (about 10 to 20%), the cause may be obscure despite exhaustive search including exploratory laparotomy (see management algorithm). Thankfully they resolve spontaneously without future problems. The rarity of adult pathology like neoplasia, diverticulitis and vascular dysplasia is reassuring. Hence the aim of management, when no cause is detectable after a reasonable search, must be to compensate for the losses and maintain stability.

Bibliography


NEWS AND NOTES

2nd WORLD CONGRESS OF PEDIATRIC GASTROENTEROLOGY, HEPATOLOGY AND NUTRITION

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PAINFUL SCROTUM

* Meiseri IV

Keyword: Acute scrotum, children, testis torsion, appendix of testis torsion

Painful scrotum is a common clinical condition in the perinatal and peripubertal period. Acute scrotum should always be explored to rule out testicular torsion. Conservative management of a torsion testis can result in necrosis within two hours. This article reviews the various differential diagnoses in child presenting with an acute scrotum.

Causes

In order of occurrence.
1. Torsion of the testis
2. Torsion of appendage of testis
3. Obstructed inguinal hernia
4. Acute hydrocele
5. Epididymoorchitis
6. Idiopathic scrotal oedema
7. Fat necrosis

Nine out of ten children with acute scrotum will have either torsion of the testis or the appendix of testis. Hence it is worth exploring rather than waiting.

Presentation of painful scrotum: It may be acute, acute on chronic or chronic. There is swelling of the scrotum with or without nausea, vomiting and fever. There may be history of similar shortlived episodes in the past. There may be history of urinary tract infection or urethral instrumentation in the past. It is very difficult to arrive at the diagnosis based on history alone. We will discuss the causes in detail separately.

Torsion of the testis (TT): History of acute pain with swelling of the scrotum with short-lived attacks in the past. There may be history of retractile testis.

On examination acute tenderness will be present with red swollen scrotum. There may be an associated hydrocele. Patient will not allow detailed examination of the involved scrotum but opposite scrotum can be easily examined. The opposite scrotum may show transverse lie of the testis, which is highly suggestive of testicular torsion. Patient needs urgent admission and exploration. Epididymoorchitis (EO) may have a very similar clinical picture except for transverse lie of the testis.

Investigations: Color Doppler Ultrasonography (CDUSG) has limited role in diagnosis. It can be done if available on the premises while waiting for the operating theatre to get ready. It is not worth getting it done on an outpatient basis. Early changes of TT and EO are indistinguishable and one may treat TT conservatively thereby endangering the testis. Isotope scan gives valuable information but is time consuming and may delay exploration.

Treatment: All children with acute scrotum should be explored at the earliest i.e. within 2 hours. Viability of testis can be judged by its colour, firmness and bleeding on incising its capsule. Portable CDUSG if available in OT is a
useful tool for assessing viability. Contralateral testis is fixed at the same sitting as the anatomy is expected to be the same. Long term follow up is necessary as atrophy of the salvaged testis is common.

**Torsion of Appendage of testis**

This is more common than TT but it is difficult to differentiate. Signs and symptoms are same as for TT. Final diagnosis is at surgery. Occasionally it may be possible to clinically differentiate, if a ‘blue dot’ sign is visible. Treatment is surgery with excision of the appendage and fixation of testis is done if it is mobile. Similar appendage on the opposite side, may be excised at the same sitting.

**Obstructed inguinal hernia**

This is a fairly common clinical condition. There may be a history of swelling which has suddenly become obstructed. There is 30% chance of neonatal hernia getting obstructed. In children the overall chance of obstruction is 20%. Hence it is better to do elective surgery at the earliest. Once obstructed, oedema of the bowel wall occurs due to vascular congestion. This leads to compromise of blood supply of the bowel leading to necrosis. It also leads to compression of testicular vessels in the cord. As this is an end artery there is a great risk of testicular necrosis.

**Signs and symptoms:** Develops as sudden onset of pain in inguinoscrotal region followed by vomiting and abdominal distension. The inguinoscrotal swelling is usually tender and irreducible. X-ray may show multiple air fluid levels with gas in the scrotum. USG confirms normal testis. Patient needs admission and conservative trial is given. This is done by keeping child nil by mouth and in the head low position with sedation and local application of ice. In 80% of the cases the hernia reduces spontaneously or by gentle manipulation. Only 20% require emergency surgery. At surgery if contents are healthy they are reduced and herniotomy done. If there is gangrene of the bowel, resection and anastomosis are done. Testis is always inspected to exclude necrosis. Once hernia is reduced spontaneously elective surgery is carried out in 48 hours time to avoid recurrent obstruction.

**Acute hydrocele**

This is difficult to distinguish from TT or obstructed hernia unless transillumination is positive. An acute hydrocele can be associated with testis torsion. Acute hydroceles are usually caused by a plug of omentum occluding the internal inguinal ring.

**Epididymoorchitis:** (Fig.2)

This is rare in children. In 100 cases of acute scrotum only 11 will have epididymoorchitis. It
may be unilateral or bilateral and may be associated with Henoch Schönlein purpura. A history of urinary tract infection, urethral instrumentation, mumps or pancreatitis needs to be elicited. Early exploration is mandatory as epididymoorchitis and testis torsion cannot be clinically differentiated.

**Rare causes**

**Idiopathic scrotal edema:**

This is rare in children. In this condition the scrotal wall is red, inflamed and thickened but the testes are painless and can be freely handled.

**Fat necrosis:**

Usually secondary to trivial trauma, felt as a small tender nodule in the subcutaneous tissue distinct from the testis. Can be treated conservatively or excised.

**Conclusion**

1. Painful scrotum is usually acute
2. Should be always treated aggressively by early surgery
3. Nine out of ten cases will be torsion testis.
4. Another 5% of children will have obstructed hernia or other causes which need exploration
5. Epididymoorchitis presenting as acute scrotum is 5% in incidence.

**Bibliography**

NEONATAL HYPERBILIRUBINEMIA-SURGEON'S PERSPECTIVE

* Archana Puri
** Yogesh Kumar Sarin

Abstract: Neonatal hyperbilirubinemia is a common problem in newborn nurseries. It manifests clinically as jaundice, which is defined as yellowish discoloration of eyes and high colored urine. Nearly 25-50% of all newborns and a much higher percentage of premature babies develop hyperbilirubinemia. This is mostly physiological, but a small percentage of these babies have pathological jaundice, requiring detailed investigation and appropriate management. This article aims to describe the diagnostic approach to neonatal hyperbilirubinemia with special emphasis on conditions, which require surgical intervention.

Keywords: neonatal jaundice, extrahepatic biliary atresia, neonatal hepatitis

Physiological versus pathological hyperbilirubinemia

In term babies physiological jaundice appears between 30-72 hours of age, reaches a peak by 4\textsuperscript{th} - 5\textsuperscript{th} day and disappears by 7-10 days of age. Total serum bilirubin does not exceed 12 mg%. Physiological jaundice in preterm babies appears earlier but not before 24 hours of age, reaches maximum intensity by 5\textsuperscript{th} – 6\textsuperscript{th} day and disappears by 8\textsuperscript{th} -14\textsuperscript{th} day. Serum bilirubin may go up to 15 mg%. Physiological jaundice can however be prolonged due to the presence of any of the following factors: 1) Immaturity, 2) Birth asphyxia, acidosis, hypothermia, hypoglycemia, 3) Drugs like salicylates, Vitamin K, frusemide, 4) Cephalhematoma and concealed hemorrhage, 5) Hypothyroidism, 6) Polycythemia and 7) Breast milk syndrome

Maisels in, 1981\textsuperscript{1} proposed criteria to exclude the diagnosis of physiological jaundice. These criteria are 1) Any jaundice which appears within first 24 hours of age, 2) Serum bilirubin concentration which increases by more than 5mg% per day, 3) Total serum bilirubin level exceeding 12 mg% in full term and 15 mg% in preterm, 4) Direct bilirubin of more than 2 mg% or conjugated bilirubin comprising more than 15% of total bilirubin level and 5) Clinical jaundice persisting for more than one week in full term and more than 2 weeks in preterm

Clinical features - When should surgical jaundice be suspected?

Clinical features, which should alert to the possibility of surgical conditions, include evidence of jaundice along with clay colored stools (absence of pigment in stools) and high colored urine\textsuperscript{3}. It is very important for physicians and surgeons to differentiate between neonatal hepatitis and extra-hepatic biliary atresia (EHBA), as the former requires conservative management and the latter needs surgical intervention at the earliest and certainly not later than 60 days of...
# Causes of Neonatal Hyperbilirubinemia

## Fig 1. Aetiology of Neonatal Hyperbilirubinemia

<table>
<thead>
<tr>
<th>Type</th>
<th>Causes</th>
<th>Subtypes</th>
</tr>
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<tbody>
<tr>
<td><strong>Medical causes</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Unconjugated</strong></td>
<td>1. Blood group incompatibility</td>
<td>ABO, Rh, minor blood group</td>
</tr>
<tr>
<td></td>
<td>2. RBC membrane defects</td>
<td>Hereditary spherocytosis, Hereditary elliptocytosis, Hereditary ovalocytosis, Hereditary poikilocytosis</td>
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<td></td>
<td>3. RBC enzyme defect</td>
<td>Glucose 6 PO₄ dehydrogenase deficiency, Pyruvate kinase deficiency</td>
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<td></td>
<td>4. Haemoglobinopathies</td>
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<td></td>
<td>5. Drug-induced hemolysis</td>
<td>Vitamin K, Sulphonamides, Antimalarials</td>
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<tr>
<td></td>
<td>7. Impaired enterohepatic circulation</td>
<td>Ileus</td>
</tr>
<tr>
<td></td>
<td>8. Increased production of bilirubin</td>
<td>Extravasated blood, Cephalhematoma, Polycythemia</td>
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<td><strong>Conjugated</strong></td>
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<td></td>
<td>2. Toxic</td>
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<td>3. Metabolic defects</td>
<td>Galactosemia, α₁-antitrypsin deficiency, Tyrosinemia, Hereditary fructoseemia</td>
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<td>4. ↑ extrahepatic circulation</td>
<td>1. Biliary atresia, 2. Paucity of intralobular bile ducts (PIBLD) - Syndromic (Alagille syndrome), - Non syndromic</td>
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<td>5. Impaired enterohepatic circulation</td>
<td>Ileus</td>
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<td></td>
<td>6. Increased production of bilirubin</td>
<td>Extravasated blood, Cephalhematoma, Polycythemia</td>
</tr>
</tbody>
</table>

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**Surgical causes** | | |
| **Unconjugated** | 1. Biliary atresia |
| | 2. Paucity of intralobular bile ducts (PIBLD) - Syndromic (Alagille syndrome), - Non syndromic |
| | 3. Choledochal cyst |
| | 4. Inspissated bile syndrome |
| | 5. Cystic fibrosis deficiency |
| | 6. Angiomatous bile duct malformation |
| | 7. Spontaneous perforation of bile duct |
| | 8. Biliary calculi |
age for better results. The clinical features are more or less similar in these conditions 1) hepatosplenomegaly, 2) ascites, 3) failure to thrive and 4) evidence of portal hypertension. Although the liver is firmer in biliary atresia and softer in neonatal hepatitis, portal hypertension is more evident in biliary atresia than neonatal hepatitis. We have to understand, at this point of time, that there are no foolproof diagnostic criteria based on clinical features to differentiate one from another. All it needs is to have a high index of clinical suspicion for EHBA in all babies with conjugated hyperbilirubinemia with acholic stools. It is prudent here to remember that 40-60% of babies with EHBA can pass normal meconium and one third of babies with severe neonatal hepatitis can pass pale stools. The specificity of pale stools for EHBA is 37.2% and sensitivity for EHBA is 87.7%.

**Diagnostic workup for surgical jaundice**

Diagnostic workup for a child suspected to have surgical jaundice is as follows:

1. Biochemical tests
   - Liver function tests
     - Bilirubin –direct/indirect
     - SGOT/ SGPT/ Alkaline phosphatase
     - Gamma glutamyl transpeptidase (GGT)
     - Prothrombin time/ concentration
     - Serum protein/ albumin
   - IgM antibody (for TORCH)
   - α1 antitrypsin level (for α1 antitrypsin deficiency)
   - Urine and serum amino acid determination
   - Urine for reducing substances (for galactosemia)
   - Bile pigments in stools
   - Urinary sulphated bile acids
   - Recent prognostic factors like hyaluronic acid, IFN-inducible protein 10 etc.

2. Duodenal juice analysis
3. Imaging investigations
   - Ultrasonography
   - Radionuclide scintigraphy (HIDA scan)
   - Magnetic resonance cholangiography
4. Endocrine investigations
   - Endoscopic retrograde cholangiopancreatography (ERCP)
5. Visual inspection of extra hepatic biliary tree
   - Laparoscopy
   - Minilaparotomy and per-operative cholangiopancreaticography
6. Liver biopsy

**Biochemical tests**

**Liver function tests (LFT):** LFT has to be done in all babies with evidence of jaundice. Total serum bilirubin along with indirect and direct bilirubin has to be measured. Conjugated hyperbilirubinemia is always pathological and merits further investigations. The liver enzymes SGOT and SGPT are elevated with hepatocyte damage and in all conditions leading to hepatocyte death; hence the non-specificity of these values. SGPT is found exclusively in liver but SGOT also occurs in the muscle, heart and kidneys. Alkaline phosphatase (ALP) is normally elevated in growing children but further elevations are indicative of biliary obstruction. ALP is also elevated in cholestasis associated with neonatal hepatitis.

Reduced serum albumin levels with prolongation of the prothrombin time (PT) are indicative of hepatocyte dysfunction and may be the forerunners to indicate hepatic insufficiency.

Gamma-glutamyl transpeptidase (GGT) is located in the hepatocyte and biliary epithelium. It will be elevated in those conditions where there is hepatocellular damage (like neonatal hepatitis) and also where there is obstruction of biliary
channels (e.g. EHBA). Karani et al\textsuperscript{4} considered that GGT is a nonspecific indicator of liver disease. However, various other authors\textsuperscript{5-7} especially Liu et al have shown a significant diagnostic accuracy of GGT in differentiating neonatal hepatitis from EHBA. In their study\textsuperscript{5} on 29 patients of EHBA and 12 patients of neonatal hepatitis before 10 weeks of age, the mean GGT values were > 600 u/l in biliary atresia but were <200 u/l in neonatal hepatitis. Its diagnostic value is further enhanced if it is used in combination with HIDA scan. GGT of >200 u/l along with non-excreting HIDA has a sensitivity of 93% and specificity of 68% for EHBA\textsuperscript{8}.

Certain special tests are needed to rule out medical causes of cholestasis. TORCH screening (IgM antibodies) should be done for all children with neonatal cholestasis. The coexistence of CMV infection with EHBA has been observed in 20-40% of EHBA patients\textsuperscript{9}. Urinary aminoacid chromatography and urine for reducing sugars to diagnose metabolic disorders like galactosemia, tyrosinemia, fructose intolerance and \(\alpha_1\) antitrypsin levels is also done.

Direct enzymatic assay of urinary sulphated bile acids can also be used in screening of patients with conjugated hyperbilirubinemia. Recently serum lipoprotein X has been proved to have a significant diagnostic accuracy in patients with EHBA. Serum lipoprotein X is positive in all patients with EHBA and in 20-40% of patients with neonatal hepatitis. It is therefore suggested that absence of lipoprotein X in serum can exclude the diagnosis of EHBA\textsuperscript{10}.

Recent research has shown certain biochemical markers in biliary atresia which predict and prognosticate hepatocellular damage. Some of these prognostic factors in EHBA are urinary D-glucaric acid (DGA) levels, serum levels of pro-collagen III peptide (P-III P), type IV collagen, plasma endothelin levels\textsuperscript{11}, serum IFM inducible protein 10\textsuperscript{12} and hyaluronic acid level\textsuperscript{13}.

**Duodenal juice analysis:** Duodenal intubation and aspiration of fluid is widely used at a few centers\textsuperscript{14,15} for differentiating EHBA from neonatal hepatitis. Ohi et al\textsuperscript{16} have shown that if duodenal juice shows the presence of bile, then EHBA is ruled out. Sensitivity and specificity were 100% and 85% respectively\textsuperscript{16}.

**Imaging investigations:** A non-invasive imaging investigation, which differentiates EHBA from neonatal hepatitis, would be the ideal choice for evaluation of conjugated neonatal hyperbilirubinemia. The various non-invasive imaging techniques available today are ultrasonography, radioscentigraphy (HIDA scan) and magnetic resonance cholangiography (MRC). Each of these investigations has its own inherent limitations, e.g., ultrasonography is operator dependant, scintigraphy has very high false positive rates and MRC is a new tool with limited experience.

Ultrasonography (USG) is the first line imaging investigation in hepatobiliary disease. It can detect a focal liver lesion of up to 5mm\textsuperscript{4}. It can also differentiate between cystic and solid lesions of liver. Ultrasonography can also pick up the intrahepatic and extrahepatic dilatation of biliary channels. Identification of gall bladder is very crucial in the diagnosis of neonatal cholestasis. In invariably all patients of neonatal hepatitis, the gall bladder is easily identified and is seen to contract after feeds. However in patients with EHBA the gall bladder is either not seen or if it is seen then it does not show any change in size with feeds\textsuperscript{17}. Recently ‘triangular cord sign’\textsuperscript{18-20} has been described in patients with EHBA. The cone shaped fibrous tissue at the porta hepatis in EHBA is seen as a triangular echogenic density on ultrasonography. This triangular cord sign is not seen in neonatal
hepatitis. Thus a triangular cord sign if seen on USG denotes EHBA but its absence does not rule it out. The positive predictive value of triangular cord sign is 95%. The other findings that can be picked up on USG are choledochal cyst, inspissated bile syndrome, biliary duct stone, or other lesions producing obstruction to bile flow.

Hepatobiliary radionuclide scintigraphy using technetium 99-m labeled iminodiacetic acid (IDA) derivatives gives an anatomical and physiological imaging of the biliary tract. The diiso-propyl compound, DISIDA, is now the preferred derivative because of its maximum concentration in the liver and low renal excretion. After the intravenous injection of DISIDA, imaging is done at 5 min. interval in the first hour and then at hourly intervals for 10 hours. If excretion to bowel is delayed, imaging is repeated at 24 hrs. In a normal subject, following isotope injection, there will be visualization of the bile ducts after 10-20 min. and the radionuclide will be seen in small bowel in 30-40 minutes. Non-excreting HIDA or DISIDA scan is usually seen in cases of obstruction to biliary flow, e.g. in EHBA. It can also be seen in severe neonatal hepatitis and in the presence of paucity of intralobular biliary ducts. In order to differentiate between EHBA and neonatal hepatitis in a non-excreting HIDA scan, baby is started on phenobarbitone (5mg/kg/day) and betamethasone (0.5mg/kg/day) for 3-5 days following which the scan is repeated. Some of the non-excretors, especially those with neonatal hepatitis can become excretors with this pretreatment. The diagnostic sensitivity, specificity and accuracy of HIDA scan in differentiating EHBA from other forms of neonatal hepatitis has been found to be 100%, 87.5% and 90.5% respectively.

Magnetic resonance cholangiography (MRC)- Wong et al reported use of MRC in jaundiced neonates and control patients. They diagnosed biliary atresia if there was non-visualization of gall bladder, CBD and CHD. This investigation, although expensive, seems promising. The distinct advantage of MRC is its ability to take images in axial, sagittal and coronal planes, with no ionizing radiation.

Endoscopic retrograde cholangiopancreatography (ERCP) was not possible in neonates and infants prior to 1980. However with the advent of pediatric side viewing endoscopes, there have been numerous reports of its use even in neonates. ERCP is especially beneficial in the diagnosis and evaluation of choledochal cyst, biliary calculi and biliary strictures in older children. Complications of ERCP such as pancreatitis, cholangitis and hemorrhage are known to occur in children as in adults. ERCP requires general anaesthesia in infants and small children, but it may be possible with heavy sedation in older children.

Visualization of extra-hepatic biliary channels

Laparoscopy: The experience of laparoscopy in the evaluation of neonatal cholestasis is very limited. The procedure can be tried in those babies where inspite of available biochemical and imaging modalities, differentiation of EHBA and neonatal hepatitis is not possible. In these cases, if one is able to visualize the extra hepatic biliary channels with normal gall bladder with bile on laparoscopy, then the possibility of EHBA is ruled out. This procedure is done under general anaesthesia and requires multiple ports for intraperitoneal access. Schier et al have found no advantage of laparoscopy over minilaparotomy with regards to anaesthesia time, morbidity, diagnostic accuracy or complications. Besides, laparoscopy has the disadvantage of being expensive to set up and is also associated with a long learning curve for the surgeon.

Minilaparotomy and per-operative cholangiogram (POC) permits the surgeon to visualize the gall bladder and extra hepatic biliary
channels. POC can be performed on table if gall bladder is patent and the egress of dye can be seen. An atretic gall bladder or non-excretion of dye into small bowel is taken as an indication for proceeding with formal dissection of the porta hepatis. If the intrahepatic and extrahepatic ducts are visualized then the cholangiopathy is secondary to cholestasis of neonatal hepatitis or inspissated bile syndrome. The POC also provides the opportunity to flush the biliary channels, which can be therapeutic for inspissated bile syndrome.

**Liver biopsy:** Liver biopsy plays an important role in the hands of an experienced pathologist, to differentiate between EHBA and neonatal hepatitis. In neonatal hepatitis, the main features are those of hepatocellular injury like lobular disarray, giant hepatocyte transformation, lobular infiltration with mononuclear cells, intracytoplasmic degeneration, relatively intact microvilli in most bile canaliculi, ballooning of hepatocytes with features of myeloid hemopoiesis (myeloid metaplasia). In EHBA however, the main features are those of bile duct damage like marked loss of bile canalicular microvilli, degenerated bile ductular cells, periductal inflammation, fibrosis and marked hepatocellular cholestasis. However, as the disease progresses, the differentiation between the pathological findings of EHBA and neonatal hepatitis gradually gets blurred. The accuracy of percutaneous liver biopsy in EHBA is approximately 82%.

Reports in literature reveal that optimal results with surgery are achieved in EHBA if surgery is done within 60 days of birth. Such cases achieve a good bile flow in 92% and a 10-year survival of 68%. In contrast to this, those babies who are operated after 90 days of life achieve good bile flow in the immediate post-operative period only in 43% and the 10-year survival is reduced to 14%. In India where pediatric liver transplant is not routinely done, the best chance of survival can only be provided if cholestatic jaundice is diagnosed early and subjected to surgery within 60 days of age.

### References


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**Summary of diagnostic workup of surgical jaundice in neonate**

<table>
<thead>
<tr>
<th>Conjugated hyperbilirubinemia</th>
<th>LFT- SGPT/ SGOT/ ALP/ PT/ Cholesterol/ S. albumin/ GGT</th>
</tr>
</thead>
<tbody>
<tr>
<td>USG</td>
<td></td>
</tr>
<tr>
<td>HIDA scan (pretreatment and post-treatment with phenobarbitone and betamethasone)</td>
<td></td>
</tr>
<tr>
<td>GB visualized</td>
<td>GB+ HIDA initially non-excretory later excretory</td>
</tr>
<tr>
<td>Excretion of HIDA</td>
<td></td>
</tr>
<tr>
<td>Conservative treatment</td>
<td>Conservative treatment</td>
</tr>
<tr>
<td></td>
<td>GB not visualized, ↑GTT HIDA non excretory</td>
</tr>
<tr>
<td></td>
<td>Minilaparotomy +POC+ liver biopsy</td>
</tr>
</tbody>
</table>


NEWS AND NOTES

IAP TASK FORCE ON CHILD AT RISK

Presents Training Programme on Management of Children in Disaster Affected Situations
In collaboration with
The Rainbow Center for International Child Health, USA and Case Western University, USA Supported by

Helping the Children Initiative, International Pediatrics Association, American Academy of Pediatrics, Johnson & Johnson Pediatrics Institute, LL/C, USA.


The workshop is intended to develop a cadre of dedicated pediatricians who would be prepared to reach disaster area anywhere in the world at a very short notice and for a period of at least 2 weeks. They are expected to be physically fit enough to live in difficult conditions.

Candidates with be selected on merit based on analysis of the information provided in the prescribed performa. Selecte candidates will have to bear their travel expenses. Course material and accommodation will be provided at a subsidized rate. Candidates who have already worked in disaster areas the proficient in emergency care will be preferred. Last date of application is 15th December 2003.

The prescribed performa will be available from
Dr. Swati Y Bhave
CII/44, Shahjahan Road, New Delhi - 110 003.
Email: sybhave@vsnl.com or from
Dr. Harish K Premde by email: harishpemde@vsnl.com

Dr. Y.C. Mathur
Chairperson

Dr. Swati Y Bhave
Convenor

IAP Task Force on Children at Risk.
ABDOMINAL MASS

* Vijayalakshmi G
** Natarajan B
*** Ramalingam A

In continuation of last issue’s masses in the abdomen let us see a few more. Fig 1 shows a mass in a thirteen year old girl arising out of the pelvis. US has shown the mass. These masses will also be seen in CT. (Fig 2). But unlike CT, ultrasound will more easily demonstrate pelvic structures like the uterus and ovaries. This is essential to find the organ of origin of the mass. To assign an ovarian origin to a mass, a normal uterus and an intact opposite ovary should be seen. But this is sometimes not possible if the tumor is too big. However it is possible to infer that a mass with such a location is most likely to be an ovarian tumor. So location is an important point in the imaging of abdominal masses. Ovarian masses can be accompanied by ascites (as in Fig 1) or peritoneal secondaries.

Fig 3 is that of a male child. There is a solid mass again arising out of the pelvis. The organ of origin in this male child was the prostate. This was a rhabdomyosarcoma. The prostatic mass compresses the posterior urethra and the child may come to you with difficulty in voiding urine. As the mass grows, it lifts the bladder superiorly and anteriorly. In advanced cases the bladder is seen in the epigastrium and there is retention of urine. CT will show the mass (Fig 4). Irregular margins and distortion of fascial planes are points in favour of malignancy. CT, and to some extent US, will show lymph nodal involvement helping in the staging of disease. The other pelvic malignant mass that you can come across is the endodermal sinus tumor.

The child in Fig. 5 came with a mass in the right iliac fossa. On general examination the child also had a small hard mass in the right calf. The plain X-ray revealed Ewing’s sarcoma in the fibula. On ultrasound the right iliac mass consisted of enlarged lymph nodes. This brings us again to the importance of a thorough clinical examination and to keep in mind the problem of secondaries.

Lymph nodal malignancy is common in the child. Fig 5 is a case of small bowel lymphoma. It shows a large hypoechoic mass with a lobulated contour. If you look carefully you can make out bowel coursing around or through the mass. Real-time US will also show peristalsis in the bowel. This tells you the plane of the mass – that it is a peritoneal mass. The mass consists of enlarged mesenteric nodes. Bowel is seen as white lines due to the air within it. These white lines run through the mass. They also block off the ultrasound so that most of the mass is obscured by the after shadow produced by gas. So careful imaging is required.

The fifteen year old girl with vague abdominal pain was sent for an ultrasound which
Fig 1 Ovarian mass. Uterus (ut). Ascitic fluid is the black area.

Fig 2 Ovarian mass - CT
Fig 3 RMS- prostate -US

Fig 4 RMS (M) seen behind the bladder (ub) -CT
Fig 5 Lymphnode secondaries.(L)

Fig 6 Lymphoma- small bowel. The white lines are bowel loops.
picked up a clover shaped thickening of the bowel. The child was advised further investigation but since there were no disturbing clinical symptoms she went away. Six months later she came with diarrhea. A repeat US was done. The same lesion had increased in size. Fig 6 shows a lobulated hypoechoic mass (m) shaped like a clover. In the centre there is a white shadow. This is the classical appearance of a bowel mass and is called the gut signature. The hypoechoic part is the irregularly thickened wall while the white echogenic area is the mucosa and represents the lumen. Colonoscopy revealed multiple colonic polyps of various sizes in the entire colon. This was a case of polyposis coli. Total colectomy was done and histopathology showed malignant change in the large polyps. Bowel masses are rare in the child. But colonic malignancies begin to appear by the end of the second decade.

In these two issues we have seen malignant solid masses in the abdomen and how to go about with a case of lump abdomen. US is the first-line investigation in the work-up of childhood abdominal malignant masses. CT scan is a secondary investigation that will help in recording extent, especially in huge masses and for complete evaluation of tumor spread. In benign childhood masses CT scan which utilizes ionizing radiation does not offer any additional benefit or information to the pediatric surgeon. We will see a few of these in the next issue.
ANTIBIOTICS IN RESPIRATORY INFECTIONS

* Balachandran A

Development of respiratory infections

The clinical spectrum of respiratory infection ranges in severity from the common cold to life-threatening illnesses such as pneumonia. Bacterial pneumonia remains a major health threat, with particular risk for infants in developing countries. The mechanisms that govern the host's response to respiratory infections are influenced by a wide variety of factors. Normally a balance exists between the host's exposure to disease-causing microorganisms and the defence mechanisms of the respiratory tract. (Fig. 1)

Upper respiratory tract infections

Upper respiratory tract infection (URI) in children presents with several signs and symptoms, of which at least one of the following should be present - cold, cough, throat pain, tenderness over sinuses, ear pain, otorrhea, mastoid swelling. On the other hand children with lower respiratory infection usually have tachypnea, chest retractions, crepitations and wheezing. Other serious conditions such as croup, epiglottitis and retropharyngeal abscess do not find a place in either URIs or LRIs. But it is wise to include them under URIs because of the lack of lower respiratory signs and absence of chest x-ray findings which are the hallmark of lower respiratory problems excluding asthma. Stridor and suprasternal retractions are indicative of upper respiratory obstruction.

URIs can be classified into three groups.

1. Benign URIs: Common cold
2. Potentially serious URIs: Group A β hemolytic streptococcus pharyngitis, Sinusitis, Otitis media, Mastoiditis.
3. Serious URIs: Croup, Epiglottitis, Retropharyngeal abscess.

What are the features suggestive of serious URI?

Respiratory distress, drooling of saliva, hoarse voice, stridor, high grade fever, systemic toxicity, altered consciousness, patch in the throat.

Lower respiratory tract infection

The causes of pneumonia depend on age, immune status, presence of underlying chronic
lung diseases. Certain infectious agents are more common at a particular age. The causative agents of pneumonia in children according to age are given in Table 1.

S.pneumoniae, H.influenzae and Staphylococcus are the bacteria that most commonly cause pneumonia in children. Mycoplasma pneumoniae and Chlamydia pneumoniae are the most common causes of “atypical” pneumonia in children. Atypical pneumonia is also called as walking pneumonia. In contrast to children with bacterial pneumonia, children with pneumonia caused by these organisms are often older than 5 years of age and the disease onset is gradual.

Though it is ideal to identify the causative microbial pathogen before starting therapy, it is practically not feasible because of contamination from upper respiratory tract flora and time required for bacteriologic culture. Considering the serious nature of the disease, the suffering children need to be started with empirical broad spectrum antimicrobial treatment immediately based on age, clinical spectrum of presentation, setting in which infection occurred, patterns of abnormality of skigram chest and the current susceptibility pattern of infecting microorganisms. Once specific organism is identified then specific narrow spectrum antimicrobial agent can be chosen. Before instituting therapy, detailed history and thorough clinical examination should be done.

Differentiating viral infection from bacterial infection is the first step in the management as it will reduce unnecessary use of antimicrobial, its cost and emergence of resistant strains (table 2). Many a time it is impossible to exclude the presence of associated bacterial infection in developing countries and the children diagnosed as pneumonia necessarily should receive antibiotics.

<table>
<thead>
<tr>
<th>Age</th>
<th>Bacteria</th>
<th>Viruses</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neonate</td>
<td>Group B streptococci, E.coli, Klebsiella sp.</td>
<td>CMV, Herpes</td>
<td>Chlamydia</td>
</tr>
<tr>
<td></td>
<td>Listeria M., S. aureus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1-3 months</td>
<td>S.pneumoniae, S.aureus, H.influenzae</td>
<td>CMV, RSV Influenza,</td>
<td>Chlamydia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Parainfluenza</td>
<td></td>
</tr>
<tr>
<td>4m-5years</td>
<td>S.pneumoniae, S.aureus, H.influenzae, Group A</td>
<td>RSV, adenovirus,</td>
<td>Mycoplasma</td>
</tr>
<tr>
<td></td>
<td>streptococcus, Klebsiella, Pseudomonas sp./</td>
<td>influenza</td>
<td></td>
</tr>
<tr>
<td></td>
<td>M.tuberculosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Over 5 years</td>
<td>S.pneumonia, S.aureus, H.influenzae, M.tuberc</td>
<td>Influenza, Varicella</td>
<td>Mycoplasma Legionella</td>
</tr>
<tr>
<td></td>
<td>ulus</td>
<td></td>
<td>sp., M.catarrhalis</td>
</tr>
</tbody>
</table>
Table 2. Clinical differentiation of pneumonia

<table>
<thead>
<tr>
<th>Feature</th>
<th>Bacterial</th>
<th>Viral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Abrupt</td>
<td>Gradual</td>
</tr>
<tr>
<td>Epidemic pattern</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Course</td>
<td>Progressive</td>
<td>Self-limiting</td>
</tr>
<tr>
<td>Temperature</td>
<td>+++</td>
<td>/ -</td>
</tr>
<tr>
<td>Toxemia</td>
<td>+++</td>
<td>-</td>
</tr>
<tr>
<td>Respiratory distress</td>
<td>++</td>
<td>+</td>
</tr>
<tr>
<td>(Infants)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Associated URTI</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Auscultation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Crackles</td>
<td>++</td>
<td>/ -</td>
</tr>
<tr>
<td>Wheeze</td>
<td>+ / -</td>
<td>++</td>
</tr>
<tr>
<td>Radiological</td>
<td>Confluent infiltrates</td>
<td>Diffuse infiltrates</td>
</tr>
<tr>
<td>(Consolidation)</td>
<td>(Consolidation)</td>
<td>in perihilar areas</td>
</tr>
<tr>
<td>Hyperinflation</td>
<td>+ / -</td>
<td>+ (RSV Infection)</td>
</tr>
<tr>
<td>Pleural involvement</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Pneumatocele</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>

In a previously normal child, presence of cough with rapid respiration is sufficient to make the diagnosis of pneumonia. The following investigations if available, like roentgenographic picture (Table 3) and CBC may provide more information.

Table 3. A guide to radiological diagnosis* of pneumonia

<table>
<thead>
<tr>
<th>Acute lobar pneumonia</th>
<th>consider pneumococcal pneumonia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right upper lobe pneumonia</td>
<td>suspect aspiration, especially in neonates and infants</td>
</tr>
<tr>
<td>Upper lobe pneumonia with cavitation</td>
<td>tuberculosis</td>
</tr>
<tr>
<td>Recurrent right middle lobe pneumonitis</td>
<td>consider partial bronchial obstruction due to glands and others</td>
</tr>
<tr>
<td>Lower lobe pneumonitis</td>
<td>chemical pneumonia</td>
</tr>
<tr>
<td>Multiple small abscesses</td>
<td>staphylococcal / klebsiella pneumonia</td>
</tr>
<tr>
<td>Severe bilateral interstitial pneumonia</td>
<td>virus</td>
</tr>
<tr>
<td>Bilateral interstitial pneumonia with malignancy</td>
<td>pneumocystis carinii</td>
</tr>
</tbody>
</table>

* The X-ray changes often lag behind clinical findings, both at the onset of pneumonia and at the time of resolution

Pneumonia - management approach

Though pneumonia can be approached and classified in different ways, classification of infectious pneumonia on the basis of presumed or proved etiology with epidemiological background is diagnostically and therapeutically more relevant and useful in day to day practice (Table 4).

Table 4. Classification of pneumonia

```
<table>
<thead>
<tr>
<th>Pneumonia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Community acquired (CAP)</td>
</tr>
<tr>
<td>Hospital acquired (HAP) or (Nosocomial)</td>
</tr>
<tr>
<td>Opportunistic (Immunocompromised) Aspiration (Anaerobic infections)</td>
</tr>
<tr>
<td>Typical</td>
</tr>
</tbody>
</table>
```
In most of the children with viral infections and atypical pneumonia syndromes there will be wheezing in addition to the other respiratory signs, which are called as “wheeze associated LRTI”. Bronchodilator therapy along with specific therapy for pneumonia will be beneficial in most of the situations.

Community acquired pneumonia (CAP)

CAP occurs as two distinct syndromes namely “Typical pneumonia syndrome” and “Atypical pneumonia syndrome”, though at times difficult to distinguish them. Since the clinical presentation, causative agents and antimicrobials used are different in the above two, prior knowledge about these two syndromes may be useful for successful management (Table 5).

CAP – Rational therapy: Since CAP is caused by variety of microbial pathogens, empirical antimicrobial therapy should be started judiciously. WHO recommends cotrimoxazole as the drug of choice in mild CAP (typical) as it is found to be cheap and effective especially if it is instituted immediately on recognition. Studies carried out in India have confirmed the efficacy of cotrimoxazole to be similar to that of ampicillin and procaine penicillin and cure rates of up to 95% have been recorded. The time taken for recovery with cotrimaxazole is similar to that with ampicillin and procaine penicillin. Cotrimoxazole is less expensive with few side effects and can be used safely by peripheral health facilities and at home by mothers. Keeping in view the safety, efficacy and the urgency for early treatment of pneumonia, cotrimoxazole has been included as one of the drugs provided to the sub-centres in the drug kit ‘A’ under the CSSM programme. Amoxycillin and second generation cephalosporins are better alternatives but cost is the limiting factor. The following table broadly outlines the spectrum of different antimicrobials in the outpatient treatment of CAP (Table 6).

<table>
<thead>
<tr>
<th></th>
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<th></th>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Cotrimoxazole</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>+ / -</td>
</tr>
<tr>
<td>Penicillin</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Ampicillin /</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Amoxycillin</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Amoxy-clav</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>I Cephalo</td>
<td>+</td>
<td>+ / -</td>
<td>+</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>II Cephalo</td>
<td>+</td>
<td>+</td>
<td>+ / -</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>III Cephalo</td>
<td>+</td>
<td>+</td>
<td>+ / -</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Macrolide</td>
<td>+ / -</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

Table 5. Differentiation of typical Vs atypical CAP

<table>
<thead>
<tr>
<th>Features</th>
<th>CAP typical</th>
<th>CAP Atypical</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Sudden</td>
<td>Gradual</td>
</tr>
<tr>
<td>Fever</td>
<td>+ + +</td>
<td>+ / -</td>
</tr>
<tr>
<td>Cough</td>
<td>Productive</td>
<td>Dry</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Pulmonary</td>
<td>Systemic</td>
</tr>
<tr>
<td>CXR</td>
<td>Localised</td>
<td>Diffuse</td>
</tr>
<tr>
<td>Organisms</td>
<td>S. Pneumonia</td>
<td>Mycoplasma</td>
</tr>
<tr>
<td></td>
<td>H. Influenza</td>
<td>Chlamydia</td>
</tr>
<tr>
<td></td>
<td>Staph. aureus</td>
<td>Legionella</td>
</tr>
</tbody>
</table>
CAP- Inpatient Management: Children with CAP who are debilitated and acutely ill, especially neonates, children with severe protein energy malnutrition, those with other system involvement, those with severe respiratory distress should be hospitalised and should be benefited with broad spectrum antibiotics based on their age, as age is an important predictor of underlying lung infection (Table 7).

**Hospital acquired pneumonia**

Hospital acquired pneumonia (HAP) is relatively more serious and complicated than CAP as children contract nosocomial infections during their hospital stay while they undergo treatment for different problems. Pneumonia acquired in institutions like hospitals or nursing homes is frequently caused by enteric gram negative bacilli (E.coli, Proteus, Klebsiella), S. aureus, P. aeruginosa or due to oral anaerobes. Empirical antimicrobials should be used based on presumptive diagnosis, as the progression of the disease is more faster than CAP (Table 8).

### Table 7. Choice of antimicrobials

<table>
<thead>
<tr>
<th>Pathogens</th>
<th>First choice</th>
<th>Alternative</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Neonatal</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>GBS, Listeria,</td>
<td>Penicillin / Ampicillin</td>
<td>Amoxiallin + Clavulanate</td>
</tr>
<tr>
<td>Gram –ve</td>
<td>Aminoglycoside</td>
<td>Cefotaxime / Ceftriaxone</td>
</tr>
<tr>
<td><strong>1-3 m</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chlamydia</td>
<td>Erythromycin</td>
<td>Co-trimoxazole</td>
</tr>
<tr>
<td><strong>3m - 5 yrs</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>S. Pneumoniae / H. Influenzae B</td>
<td>Penicillin / Amoxicillin / Co-amoxiclav, Cefuroxime</td>
<td>Cefotaxime</td>
</tr>
<tr>
<td>S. aureus</td>
<td>Cloxacillin</td>
<td>Ceftriaxone</td>
</tr>
<tr>
<td><strong>5 yrs &amp; above</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mycoplasma</td>
<td>Erythromycin</td>
<td>Vancomycin</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Clarithromycin / Azithromycin</td>
</tr>
</tbody>
</table>

### Table 8. Hospital acquired pneumonia: Empirical therapy

<table>
<thead>
<tr>
<th>Presumptive pathogen</th>
<th>Suggestive antimicrobials</th>
</tr>
</thead>
<tbody>
<tr>
<td>S. aureus</td>
<td>Cloxacillin/Vancomycin</td>
</tr>
<tr>
<td>Gram (-) bacilli</td>
<td>Aminoglycosides</td>
</tr>
<tr>
<td>E-coli, Klebsiella</td>
<td>Ticarcillin + Clavulanate</td>
</tr>
<tr>
<td>P. aeruginosa</td>
<td>Ceftazidime / Imipenem</td>
</tr>
<tr>
<td>Anaerobes</td>
<td>Metronidazole, Clindamycin</td>
</tr>
<tr>
<td></td>
<td>Cefoxitin, Penicillin</td>
</tr>
</tbody>
</table>

### Opportunistic pneumonia

Opportunistic microbes are those with minimum pathogenic potential or non-pathogenic, may become opportunistic pathogens and cause disease exclusively in compromised hosts with defect in the normal defense mechanism or on immunosuppressive therapy. Lower respiratory tract serves as an important portal entry for systemic infection and constitute 25% or of total
infections in such patients. Pneumocystis carinii and other fungi such as Aspergillus fumigatus, Viral infections (RSV, Herpes simplex, CMV, Influenza) and Mycobacterium tuberculosis are the common infections encountered. Since sputum may be contaminated with upper respiratory flora, bronchoalveolar lavage (BAL) obtained by fiberoptic bronchoscopy play a significant role in isolation of offending organism (Table 9).

Table 9. Immunocompromised children - Pathogens

<table>
<thead>
<tr>
<th>* Bacteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gram +ve : S. pneumoniae, S. aureus, M. tuberculosis</td>
</tr>
<tr>
<td>Gram –ve : Pseudomonas, H. influenzae b, Legionella</td>
</tr>
<tr>
<td>* Viral</td>
</tr>
<tr>
<td>RSV, Herpes simplex, CMV, HIV, Varicella</td>
</tr>
<tr>
<td>* Fungal</td>
</tr>
<tr>
<td>Pulmonary aspergillosis</td>
</tr>
<tr>
<td>* Parasitic</td>
</tr>
<tr>
<td>Pneumocystis carinii</td>
</tr>
</tbody>
</table>

Since respiratory infections constitute more than one third of the patients in hospital and office practice, each physician should have indepth knowledge both about causative organisms and successful management with rational antimicrobial therapy. The art of choosing appropriate antimicrobial therapy against different respiratory infections is the secret behind the success of eminent clinicians who invariably score over others in clinical practice.

References


NEWS AND NOTES

8TH ASIAN AND OCESANIAN CONGRESS OF CHILD NEUROLOGY
October 7-10, 2004

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SLEEP: A DYNAMIC ACTIVITY

* Kamlesh Lala

Sleep is a state of unconsciousness from which a person can be aroused by sensory or other stimuli. Until the 1950s, sleep was thought as a passive, dormant part of a life. But it is now known that brain is very active during sleep and it affects daily functioning and physical and mental health of a child in many ways.

Neurotransmitters such as serotonin, melatonin and norepinephrine control whether a child is asleep or awake by acting on different neurons in the brainstem. Adenosine builds up in the blood while a child is awake and causes drowsiness.

Stages of sleep

During sleep, the child usually passes several times through four stages of non rapid eye movements (NREM) and rapid eye movement (REM) sleep in that order. Child spends almost 50% of his total sleep time in stage 2, about 20% in REM sleep and the remaining 30% in the other stages. Premature babies spend about 80% of their time in REM sleep while it is only 50% in full term infants. It decreases to 1.5 to 2 hours when the child reaches puberty.

During stage 1, which is light sleep, children drift in and out of sleep and can be awakened easily. Eye movement and muscle activity are slower. Child awakened from stage 1 sleep often remembers fragmented visual images and may experience sudden muscle contractions called hypnic myoclonia, often preceded by a sensation of starting to fall. During stage 2 sleep, the eye movements stop and brain waves become slower, with occasional bursts of rapid waves called sleep spindles. In stage 3, extremely slow theta waves begin to appear, interspersed with smaller, faster waves. By stage 4, the brain produces delta waves almost exclusively. It is very difficult to wake a child up during stages 3 and 4, which together are called deep sleep. There is no eye movement or muscle activity. Children awakened during deep sleep do not adjust immediately and often feel groggy and disoriented for several minutes after they wake up.

During REM sleep, breathing becomes more rapid, irregular and shallow, eyes jerk rapidly in various directions and limb muscles become temporarily paralyzed. Heart rate increases, blood pressure rises and males develop penile erections. When child awaken during REM sleep, he often describes bizarre and illogical tales - dreams.

The first REM sleep period usually occurs about 70 to 90 minutes after child falls asleep. A complete sleep cycle takes 90 to 110 minutes on average. The first sleep cycle each night contain relatively short REM periods and long periods of deep sleep. As the night progresses, REM sleep periods increase in length while deep sleep decreases. By morning, child spends nearly all his sleep time in stages 1, 2 and REM.

Factors affecting sleep

Since sleep and wakefulness are influenced by different neurotransmitter signals in the brain, foods and medicines that change the balance of

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these signals affect the sleep pattern. CNS stimulants like caffeine and decongestants may cause insomnia.

Children lose some of the ability to regulate their body temperature during REM sleep. So abnormally hot or cold temperatures in the environment can disrupt this stage of sleep. If REM sleep is disrupted one night, body don’t follow the normal sleep cycle progression the next time and child often slips directly into REM sleep and goes through extended periods of REM.

Necessity of sleep

As of today it is not clear, why we need sleep, but animal studies show that sleep is necessary for survival. Sleep-deprived rats survived less and developed abnormally low body temperatures and sores on their tail and paws because of impaired immune system.

Deep sleep coincides with the release of growth hormone in children and young adults. Many of the body’s cells also show increased production and reduced breakdown of proteins during deep sleep. Activity in parts of the brain that control emotions, decision-making processes and social interactions is drastically reduced during deep sleep, suggesting that this type of sleep may help children maintain optimal emotional and social functioning while they are awake.

REM sleep stimulates the brain regions used in learning. This may be important for normal brain development during infancy, which would explain why infants spend much more time in REM sleep than adults. Like deep sleep, REM sleep is associated with increased production of proteins.

Duration of sleep needed

The amount of sleep each child needs depends on many factors, including age. Infants generally require about 16 hours a day, while teenagers need about nine hours on an average. For most adults the optimum requirement is only 6 to 8 hours. The amount of sleep a child needs also increases if he has been deprived of sleep in previous days. It is said that optimum requirement of sleep is “as much as needed to feel refreshed and healthy the next day, but no more”.

Sleep deprivation

If a child feels drowsy during the day, even during interesting activities and if he routinely falls asleep within five minutes of lying down, he probably has severe sleep deprivation. Microsleeps, or very brief episodes of sleep in an otherwise awake child, are another mark of sleep deprivation. The drowsiness caused by sleep deprivation is not affected by CNS stimulants.

Many studies make it clear that sleep deprivation is dangerous. Too little sleep leaves a child drowsy and unable to concentrate the next day. It leads to impaired memory and physical performance and reduced ability to carry out math calculations. If sleep deprivation continues, children may become irritable. Without sleep, neurons may become so depleted in energy or so polluted with byproducts of normal cellular activities that they begin to malfunction.

Dreams

Children typically spend more than two hours each night dreaming. How or why we dream is still a hot topic of research. Sigmund Freud, believed dreaming was a “safety valve” for unconscious desires.

Only after 1953, when researchers first described REM in sleeping infants, did scientists begin to carefully study sleep and dreaming and concluded that dreams almost always occur during REM sleep. REM sleep begins with signals from pons at the base of the brain. These signals travel to the thalamus, which relays them to the cerebral
cortex—that is responsible for learning, thinking, and organizing information. The pons also sends signals that shut off neurons in the spinal cord, causing temporary paralysis of the limb muscles. If something interferes with this paralysis, children will begin to physically “act out” their dreams—a rare, dangerous problem called REM sleep behavior disorder. A child dreaming about a ball game, for example, may run headlong into furniture or blindly strike someone sleeping nearby while trying to catch a ball in the dream.

Some believe dreams to be the cortex’s attempt to find meaning in the random signals that it receives during REM sleep creating a “story” out of fragmented brain activity.

**Sleep and circadian rhythms**

Circadian rhythms are regular changes in mental and physical characteristics that occur in the course of a day (circadian is Latin for “around a day”). Most circadian rhythms are controlled by the body’s biological “clock” — the suprachiasmatic nucleus (SCN) in the hypothalamus. Light signals from the retina travel along the optic nerve to the SCN and then to the pineal gland, which responds to light-induced signals by switching off production of melatonin. The body’s level of melatonin normally increases with darkness making people feel drowsy. The SCN also governs functions that are synchronized with the sleep/wake cycle, including body temperature, hormone secretion, urine production and changes in blood pressure.

Circadian rhythms can be affected to some degree by almost any kind of external time cue, called zeitgebers (German for “time givers”) such as the beeping of alarm clock or the timing of meals.

The children with disturbed circadian rhythm have an increased risk of cardiac problems, digestive disturbances and emotional and mental problems, all of which may be related to their sleeping problems. Many children with total blindness experience life-long sleeping problems because their retinas are unable to detect light. These children have periodic insomnia. Daily supplements of melatonin may improve nighttime sleep for such patients but not advisable for long term use.

**Sleep and disease**

Sleep and sleep-related problems play a role in a large number of disorders and affect almost every field of medicine. Asthma and epilepsy have relation with sleep. For example, asthmatic attacks tend to occur more frequently during the night and early morning and sleep deprivation triggers seizures.

Neurons that control sleep interact closely with the immune system. Infectious diseases tend to make a child feel sleepy because cytokines are powerful sleep-inducing chemicals. Sleep may help the body conserve energy and other resources that the immune system needs to mount an attack.

Sleeping problems occur in almost all children with mental disorders. Children with depression, for example, often awaken in the early hours of the morning and find them unable to get back to sleep. Sleep deprivation may decrease the pain threshold.

**Sleep disorders**

Though there are number of sleep disorders described (Table 1) only few are common among the children. Few of these are discussed below.
Table 1. International classification of sleep disorders in adolescents

1. Dysomnias. Problems associated with the onset and maintenance of sleep
   a. Intrinsic dysomnias
      i. Obstructive sleep apnoea
      ii. Narcolepsy
      iii. Restless leg syndrome
      iv. Alveolar hypoventilation
   b. Extrinsic dysomnias
      i. Difficulty with sleep onset
      ii. Difficulty with sleep hygiene
      iii. Limit setting difficulty
      iv. Excessive intake of food, drugs, medicines etc.
   c. Alteration in circadian rhythm
      i. Difficulty due to time schedule
      ii. To early or too late sleep onset
      iii. Inadequate or variable time schedule

2. Parasomnias. Phenomena that occur during the period of sleep
   a. Disorders of awakening
      i. Confusional awakening
      ii. Somnambulism
   b. Problems during the transition awake-sleep
      i. Rhythmic movements at sleep onset
      ii. Jactano Capitis Nocturna
   c. Disturbances during REM sleep
      i. Nightmares

3. Other parasomnias
   a. Enuresis
   b. Night eating syndrome
   c. Nocturnal paroxysmal dystonia

Night terrors: Night terrors are a different phenomenon from nightmares and can be quite upsetting for a parent to watch. About 90 to 180 minutes after falling asleep, the youngster will abruptly sit up in bed, open his eyes and scream loudly or cry out for help. For the next few minutes he may gasp, moan, mumble, thrash about and seem to be in a confused, agitated state. His breathing and heart rate will accelerate significantly. He will be unresponsive to his parents’ attempts at comforting him and may even push them away. These episodes can sometimes last for 30 to 60 minutes before the child rather quickly returns to a peaceful sleep, remembering nothing about it the next morning and leaving parents baffled and terrorized - hence, the name “night terrors.”

Night terrors (pavor nocturnus) occur in a relatively small number of children (1 to 5%), taking place during a nondreaming, deep stage of sleep. As frightening as they may be for parents, they are not a reflection of a psychological disturbance. They are a normal, although infrequent, part of the body’s transition between sleep states. Sometimes physical exhaustion can contribute to a child’s night terrors. Most children outgrow night terrors without treatment and parents can do nothing to resolve their occurrence. Parental patience and understanding are important, as these night terrors tend to be much more stressful for parents than for their children.

Nightmares: Nightmares are common in middle childhood. In a typical episode a child will have a scary dream, filled with monsters or other frightening beings. He may awaken, become anxious, breathe heavily and begin crying, may resist going back to sleep, needing close and constant reassurance.

In most children nightmares occur only occasionally. It usually occurs in REM stage. If they happen often - or if the same frightening dream recurs - evaluate the child for stressful conditions.

Sleep talking (somniloquy): Sleep talking occurs more often than sleepwalking. During sleep, the child begins speaking, often unintelligibly and in a monotonous voice and usually for no more
than thirty seconds. Most episodes take place during nondreaming sleep. Treatment is rarely needed.

**Sleepwalking (somnambulism):** About 15% of all children aged between 5 and 12 have at least one sleepwalking episode. Sleepwalking usually occurs during the second or third hour of nighttime sleep and may occur several nights a week, being more common in boys than in girls. The child sits up and without totally awakening, leaves his bed, usually walking awkwardly, with his eyes open and a blank look on his face. For several minutes he may wander through the house, even opens doors along the way, but his actions are purposeless. If spoken to, he may seem to respond, but the words are usually unintelligible. He will probably return to his bed on his own and go back to normal sleeping, recalling nothing of this nighttime activity when he awakens in the morning.

Management requires ensuring safe environment e.g. lock the outside doors, block the stairways etc. Sleepwalking tends to run in families. In most children this problem disappears on its own, generally by early adolescence.

**Restless legs syndrome:** Restless legs syndrome (RLS), a familial disorder causing unpleasant crawling, prickling or tingling sensations in the legs and feet and an urge to move them for relief, is emerging as one of the most common sleep disorders especially among older children. Many RLS patients also have a disorder known as periodic limb movement disorder (PLMD), which causes repetitive jerking movements of the limbs, especially the legs. These movements occur every 20 seconds to 40 seconds and cause repeated awakening and severely fragmented sleep.

Drugs that affect the neurotransmitter dopamine often can relieve RLS and PLMD, suggesting that dopamine abnormalities underlie symptoms of these disorders'. Understanding how these disorders occur may lead to better therapies in the future.

**Sleep apnea:** This is a rare condition seen in children. Sleep apnea is a disorder of interrupted breathing during sleep. It usually occurs in association with obesity, enlarged adenoids and tonsils or loss of muscle tone. These changes allow the airway to collapse during breathing when muscles relax during sleep. This problem, called obstructive sleep apnea (OSA), is usually associated with loud snoring (though not everyone who snores has this disorder).

During an episode of obstructive apnea the airflow is blocked for 10 seconds to a minute while the sleeping child struggles to breathe. When his blood oxygen level falls, the brain responds by awakening him enough to tighten the upper airway muscles and open the airway. The child may snore or gasp, then resume snoring. This cycle may be repeated hundreds of times a night. Because of frequent awakening the child may suffer from sleep deprivation. Sleep apnea also deprives the child of oxygen, which can lead to morning headaches or a decline in mental functioning. Patients with the typical features of sleep apnea, such as loud snoring, obesity, and excessive daytime sleepiness must be screened for OSA. It is best diagnosed by polysomnography. This test records the patient’s brain waves, heartbeat, and breathing during an entire night. Mild sleep apnea frequently can be overcome through weight loss or by preventing the child from sleeping on his back (tying a tennis ball on the back). Others may need special devices or surgery (uvulopalatopharyngoplasty) to correct the obstruction. Children with sleep apnea should never be given sedatives as it can prevent them from awakening enough to breathe.

**Narcolepsy:** This is also a rare condition seen in children. Children with narcolepsy have frequent repetitive “sleep attacks” lasting from
several seconds to more than 30 minutes at various times of the day, even if they have had a normal amount of nighttime sleep. They tend to fall asleep when unstimulated or carrying out monotonous activities like bathing, eating. Children with narcolepsy also may experience cataplexy (loss of muscle control during emotional situations), hypnogogic hallucinations and temporary paralysis. When this occurs, muscle relax and they may fall to the floor. The symptoms of narcolepsy typically appear during adolescence, though it often takes years to obtain a correct diagnosis. The disorder (or at least a predisposition to it) is usually hereditary, but it occasionally is linked to brain damage from a head injury or neurological disease. Once narcolepsy is diagnosed, stimulants, antidepressants or other drugs can help control the symptoms and prevent the embarrassing and dangerous effects of falling asleep at improper times. Naps at certain times of the day also may reduce the excessive daytime sleepiness. Recently a gene has been identified that causes narcolepsy - a breakthrough that brings a cure for this disabling condition within reach.

The Future

Sleep research is expanding and attracting more and more attention from scientists. Sleep is an active and dynamic state that greatly influences our waking hours. Innovative techniques, such as brain imaging, can now help researchers understand how different brain regions function during sleep and how different activities and disorders affect sleep. Understanding the factors that affect sleep in health and disease also may lead to revolutionary new therapies for sleep disorders and to ways of overcoming jet lag and the problems associated with shift work.

Bibliography


NEWS AND NOTES

SNIDs AND NIDs FOR POLIO – 2003-2004

Government of India has organized Subnational Immunization days (SNIDs) on 14th September 2003 and 9th November 2003 and National immunization days (NIDs) on 4th January 2004 and 22nd February 2004 for polio immunization. SNIDs will be conducted all over the state in the states of Uttar Pradesh, Bihar, Delhi, Gujarat, Haryana, Rajasthan, Jharkhand and West Bengal and in 31 districts of MP and 4 districts of Uttaranchal. NIDs will be conducted all over India. All IAP members are requested to keep their clinics open and advice parents to bring their children on these dates for polio immunization.

T. Jacob John Naveen Thacker
Chairperson Convener
Polio Eradication Committee, IAP
MAINTENANCE OF INHALATION DEVICES

*Nagabhushana S*

Inhalation therapy is the corner stone of successful management of both acute and long term care of childhood asthma and a host of other childhood respiratory illnesses\(^1,2\). With increasing levels of awareness about the benefits of inhalation therapy and the lack of significant side effects, the use of these equipments has increased worldwide.

A proper understanding regarding the use and maintenance of these equipments is necessary to use them efficiently\(^2,3\).

**Metered Dose Inhalers (MDI)**: They are the most common devices used in ambulatory practice. It is always preferable to use them attached to spacer devices to avoid hand lung incoordination. The following are the Do’s and Don’t’s about MDI.

**DO’S**
- Store below 30 degrees
- Keep away from heat and direct sunlight
- Keep away from children
- If the inhaler becomes very cold, remove canister and warm it in your hands for a few minutes. Do not use other forms of heat.
- Always keep a ‘Puff Count’
- Always keep the nozzle of the MDI clean.

**DON’T’s**
- Do not puncture the canister or incinerate.
- Avoid excess heat.
- Do not freeze.
- Do not tamper with valve.
- Do not use after the date of expiry.
- Do not interchange a reliever for a preventer.

**Note**: A full canister when held in the hand appear relatively heavy and sinks to the bottom of the container filled with water. A canister which is 3/4 empty may not always deliver the appropriate dose of the drug. It may either be discarded or the dose doubled.

**Cleansing**:
- Clean your inhaler atleast once a week, pull the metallic canister out of the box of the inhaler. Remove the mouth piece cover, rinse the body and mouth piece in warm water, leave it to dry in a warm place, replace the canister and mouth piece correctly.

**Spacers**:
- It is preferable to use a spacer with a well fitting soft face mask in young children and toddlers. Spacers with reliever medications attached can be used to successfully treat an acute episode of asthma with an efficacy similar to nebuliser, especially if the tidal breathing technique is adopted. Further spacers are easy to clean, more portable, less expensive and are convenient to use\(^4,5,6\).

**Cleansing**:
- Wash your spacer with a mild soap/detergent solution every week.
- Allow it to drip dry.

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Do not rinse or use a cloth to wipe it dry (initiates static charge which prevents proper drug delivery).

**Maintenance**

- Change your spacer if the valve or body is not functioning or is damaged.
- After cleansing, each time ‘prime’ the spacer with a dose of drugs used.
- Change your spacer at least once every two years.
- Prefer small volume spacers for infants and toddlers who have significantly small tidal volumes.

**Rotahaler**

- Keep the equipment clean and dry
- Keep rotahaler away from dampness.
- If the rotacap does not open after insertion:
  - Loosen the capsule manually or
  - Cut the upper 1/3 with a sharp blade and reinsert
- Clean the inside of the rotahaler with a brush twice a week. Rinse all parts of the rotahaler in warm water and dry it in a warm place.

**Nebulizer**

Which nebulizer to buy? 7, 8, 9, 10, 11

It is preferable to buy a piston driver jet nebulizer with a longer nebulisation time rather than a diaphragm driver nebuliser which creates more sound and is less durable though less expensive.

The nebuliser should have a facility to administer oxygen simultaneously which is useful during acute severe wheezing to correct hypoxia which is often co-existent.

Arguably, an ideal nebuliser should have a high drug output (respirable fraction of at least 50% of their driven gas flow), a small droplet size equally during inspiration and expiration for penetration into the lung and a short nebulisation time for patient compliance.

Water soluble drugs like Beta2 agonists are easily and rapidly nebulised (around five minutes) while lipophilic drugs, eg: Inhaled steroids are slowly (around ten minutes) and incompletely nebulised.

**Practical points** 1, 7, 9

Saline should be used as a diluent and not distilled water, as water can cause reflex bronchospasm.

Ensure a gap of few minutes between two doses of medications.

Preferably nebulise in the upright position

- Do not exceed the ‘fill in’ volume of 4 ml.
- Always use a soft well fitting tight facemask for optimal drug delivery
- A mouth piece is preferred in older children
- Optimal nebulisation time is usually five minutes.
- Do not mix two medications but nebulise them separately (a possible exception being salbutamol with Ipratropium or cromoglycate)
- A period of rest of five minutes to the equipment is necessary if the nebuliser has run for five minutes.

Tapping the nebuliser chamber when the solution begins to “splutter” increases the volume of the output.

**Maintenance** After each use 4, 9: Cleanse thoroughly the tubing, chamber, etc. with running...
tap water. Cleanse mask with a mild disinfectant and ‘dry run’ the nebulizer for a minute.

After each day:

· Disassemble the nebuliser completely
· Submerge tubing, medication cup and mouth piece/masks in a mild liquid detergent and warm water for a few minutes.
· Using a small bristle brush, scrub off any sedimentation on any of the parts.
· Rinse all parts after washing to remove detergent
· Immerse all parts with acetic acid disinfectant solution for 10 minutes. (Acetic acid solution is prepared by mixing one part of Vinegar with three parts of water prepared fresh every day).
· The housing can be cleaned with a slightly damp cloth.
· Never spray any liquid into compressor housing.

In our environment, it is advisable to keep the top of the nebuliser and the mouthpiece covered with dust caps and stored in a clean area.

Replacement:

· Change the filter after 250 hours of use or earlier depending on the change in the color of the filter.
· The compressor needs calibration / overhauling atleast once a year.

Nasal spray:

Wipe the nozzle(applicator) with a clean cloth or tissue after every use, and replace the dust cap.

Nasal applicator should be cleansed once a week. After removing the dust cap off, pull the actuator upwards very gently to detach it from the cap. Soak the actuator and dustcap in warm water for a few minutes and rinse under running tap water. Shake off the excess water and allow it to dripdry. Refit the actuator(nasal applicator) very carefully and gently.

Do not try to clean the spray hole with a pin or sharp object because this will destroy the spray mechanism.

Note: With all inhalation devices, a brief period of breath holding after inhalation provides better drug deposition in airways.

Patient education

For proper use of any form of inhalation device by the child, the corner stone of success is the proper understanding and knowledge about the equipment by the doctor as well as adequate time spent with the child and his parents to explain and demonstrate the proper technique and maintenance of the device. This needs to be reinforced during subsequent visits12, 13, 14

Proper patient education ensures both confidence and compliance on the part of the child as a successful partner in using inhalation therapy optimally14.

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**BOOK REVIEW**

**Title** : Clinical Pediatrics  
**Author** : Dr. B. Anjaiah  
**Review** : The 2nd edition of the Text Book Clinical Pediatrics is a concise coverage of important topics in the various subspecialties in pediatrics. The author has selected the various issues meticulously and has covered in depth some of the topics. This book is “Reader Friendly” and includes the recent advances in the various specialties. This book covers relevant problems in cardiology, neurology, nephrology, gastroenterology and respiratory medicines. The section in pediatric surgery is also well represented. The author has included some problems in neonatology also. This book does not include many of the infectious diseases. It is a hardy book, which can be beneficial both to the medical students undergraduates or postgraduates as well as medical practitioners.

**Publisher** : Divyesh A Kothari  
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Hyderabad – 500 095.  
Andhra Pradesh.

**Price** : Rs.450/-
AN UNUSUAL CAUSE OF RECURRENT G.I. BLEED

* Malathi Sathiyasekaran

A 5 Year-old boy presented with history of intermittent hematemesis and melena of two weeks duration, during which period he required two blood transfusions. The bleed was preceded by colicky abdominal pain. There was a history of fall from a tricycle three weeks ago. Two days after the fall he developed jaundice, which subsided spontaneously. There was no past history of ascites, pedal oedema or bleeding diathesis.

On examination, he was pale, undernourished and weighed 13kg. There was no icterus or lymphadenopathy. Abdomen was slightly distended, but soft. Liver was palpable 2.5cm below the right costal margin. The bowel sounds were normal. A clinical diagnosis of hemobilia based on the triad of GI bleed, abdominal colic and jaundice was made.

Investigations revealed hemoglobin of 9.6gm/dl, PCV 30%, platelet count 6.02 L/mm³ and a normal white count. His bilirubin was 0.7mg/dl, with a direct component of 0.3mg/dl, SGOT 73 U/L, SGPT 58 U/L, SAP 600U/L. The bleeding time was 2 minutes, prothrombin time was 14sec and control 13sec, activated plasma thromboplastin time was 28sec and control 30sec. Chest skiagram and upper GI endoscopy were normal. Ultrasonography of the abdomen revealed a 3.4 x 2.8cm hypoechoic area in the right lobe of liver with a prominent bile duct near it.

Doppler study of the abdomen revealed a well-defined hematoma, sized 4.2 x 3.7 x 2.9cm close to the portal bifurcation, with a mildly dilated biliary radicle adjacent to the hematoma. A small fluid filled cavity of 1cm diameter was also seen between the hematoma and portal bifurcation, which filled with blood and showed an arterial flow pattern similar to the hepatic artery (Fig- 1). The findings were suggestive of a well-defined hematoma in the right lobe of liver with probable pseudoaneurysm adjacent to and communicating with the right hepatic artery.

Interventional radiology and surgical options were contemplated, but he responded well to conservative management with blood transfusion, intravenous fluids and antibiotics and there was no further bleed. Ultrasound was repeated and it showed a significant decrease in the size of the lesion.

The term hemobilia coined by Sandblom in 1948 means “bleeding into the biliary tract” and is due to a communication between a blood vessel and the bile duct. The communication may be intrahepatic or extrahepatic. In 1871 Quinke described the course of events in a biliary tract bleed and established the three cardinal symptoms of GI bleed, biliary colic and jaundice as “Quinke’s triad”. The diagnosis of hemobilia is often missed or delayed because of a low index of clinical suspicion and non-availability of investigation techniques such as Doppler ultrasound, CT scanning and angiography. In a study of obscure GI bleed it constituted 2.5% of the total cases¹.
The etiology of hemobilia can be varied such as accidental trauma, iatrogenic following procedures like liver biopsy or percutaneous transhepatic cholangiogram (PTC), gallstones, tumors, biliary tract infections by bacteria or parasites, liver abscesses, pancreatitis and vascular malformation. Tuberculosis has also been reported as a cause in India.²

Accidental trauma leading to hemobilia occurred in 38.6% of Sandblom’s series whereas iatrogenic cause constituted 65% in the reports by Green et al³. The pathogenesis of trauma causing hemobilia is due to central liver rupture without capsular tear. The formation of a hematoma within the liver results in bile stasis. The static bile impairs healing and the necrotic tissue erodes into a vessel creating a fistula. The presentation may be acute or delayed. Hemobilia can also arise from a larger cavity into which both the blood vessels and bile ducts open.

The sequences of events in hemobilia differ depending on whether the bleed is profuse or slow. In profuse bleeding the clot formed is mixed with bile. The fibrinolytic activity of the bile dissolves the clots, which therefore adhere to the mucosa of the bile duct. If the bleeding is slow a pure cylindrical blood clot taking the shape of the bile duct lumen is formed. The bile can also lyse these clots.

The clinical manifestations of hemobilia are therefore related to the rapidity of bleeding, amount of blood loss and presence of blood and clots within the biliary tree. The bleed can manifest either as hematemesis or melena or both. The bleeding can be slow, intermittent and present as recurrent melena. The presence of blood or retained clot within the biliary line can cause biliary colic, jaundice or cholangitis.

The diagnosis of hemobilia may be apparent if the triad of bleed, jaundice and abdomen colic is present as seen in this child. However, this triad is seen only in 20% of cases².

Ultrasound scan with color doppler is a very sensitive, non-invasive investigation to confirm the diagnosis. Upper gastrointestinal endoscopy may demonstrate bleeding from the ampulla of Vater. This will be evident only in 12% of cases.⁴

Fig 1. Hematobilia with arterial flow pattern similar to the hepatic artery
Computerized sonography with contrast is a useful test if color doppler is not able to delineate the aneurysm. ERCP / MRCP are done only if the above investigations are not conclusive. Selective hepatic artery angiogram is an ideal method to demonstrate the site of bleeding and also helps in arterial embolization of the bleeding vessel.

The therapeutic options to treat hemobilia depend on the clinical condition of the patient, severity of the bleeding, the site (intrahepatic / extrahepatic) and the type of pathology and the available expertise. The commonly employed options include angiographic embolization, hepatic arterial ligation and liver resection. In accidental trauma if hemobilia is associated with hemoperitoneum, exploratory laparotomy is done and hemostasis achieved. If patient’s condition does not warrant surgery, hemobilia can be managed conservatively about 7% of patients can be expected to recover spontaneously.

**Key message:**
- Hemobilia is an unusual cause of GI bleed in a child but should be considered whenever a child presents with abdominal pain, GI bleed with or without jaundice.
- A history of trauma may not be obtained initially as the presentation may be delayed.
- Ultrasound color doppler is an ideal noninvasive tool in diagnosis.
- If the bleed continues, interventional radiology if available is an option; otherwise surgery may be the method of management.

**References**


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**NEWS AND NOTES**

**MEDICARE INDIA 2004**

April 6-8, 2004

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WILLIAMS’ SYNDROME

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Williams’ syndrome is an uncommon genetic disorder associated with characteristic facies, supravalvular aortic stenosis, peripheral pulmonary stenosis, mental retardation, hypertension, and premature aging of skin. We present this child who was referred to us for the evaluation of a cardiac murmur. The characteristic facies and personality led us to suspect this syndrome; the cardiac lesions were confirmed by echocardiography and catheterization, which corroborates Williams’ syndrome. We focus on the cardiac workup and discuss the various cardiovascular manifestations of this Syndrome.

A 4½ year-old male child born of a third degree consanguinous parentage presented to us with a cardiac murmur detected on a routine health check up. He was the second of three siblings born of a full term normal delivery. The child had no specific cardiac symptoms.

The clinical examination revealed an active, playful, talkative mentally retarded child. The dysmorphic facies included a small chin, short philtrum, blunt nose, depressed nasal bridge, medial epicanthal fold, hypertelorism with dental malformation and malocclusion (Figure 1). He was acyanotic and had no clubbing or anemia.

The pulse was regular at 106/ minute and there was no radiofemoral delay. Carotid and suprasternal thrill were present. The blood pressure recorded in the right upper limb was 130/70 mmHg and left upper limb was 90/70 mmHg. The JVP was normal.

The examination of the cardiovascular system revealed a heaving apical impulse in the 4th left intercostal space on the midclavicular line. The first and second heart sounds were normal and there were no added sounds. A grade 3/6 ejection systolic murmur was heard best in the right 2nd intercostal space and conducted to the carotids.

Screening for renal, electrolyte abnormalities, serum calcium, routine chemistries and hemogram were all within normal limits. The ECG and Chest X-ray were within normal limits. Transthoracic 2D echocardiography detected left ventricular hypertrophy and colour doppler detected turbulence in the aortic root. Continuous wave doppler interrogation estimated the supravalvular aortic stenosis gradient to be 55 mmHg.

Cardiac catheterization demonstrated pull-back pressure gradient of 35 mm Hg across the supravalvular aortic stenosis and left ventriculography demonstrated hour-glass type of supravalvular aortic stenosis. Right heart catheterization revealed gradient of 5 mmHg between main and right pulmonary artery (RPA) and pulmonary arteriogram demonstrated stenosis at the origin of the right pulmonary artery.

Outflow obstruction with gradient more than 40mmHg is generally considered significant enough to warrant intervention. As the doppler measured gradient is instantaneous, it is more physiological than catheter pull-back gradient.
obtained in the catheterisation lab. This explains the discrepancy in the stenosis gradients by both methods. In ambiguous situations, simultaneous recording of pressures proximal and distal to the aortic stenosis will provide accurate gradient. The presence of significant supravalvular aortic stenosis in our patient may necessitate intervention at a later date. RPA stenosis does not warrant intervention as the gradient is insignificant.

Patient’s family deferred surgery after discussion of risks and benefits and currently he is well after 10 months of follow-up without decompensation. At this time we are following the patient with serial echocardiograms annually to diagnose development of left ventricular dysfunction and coronary ectasia secondary to high pressure proximal to the stenosis. These would also warrant early surgical intervention. Apart from infective endocarditis prophylaxis and general attention to health and oral hygiene, no specific cardiac medications are indicated.

Discussion

Williams’ Syndrome was first described by Fanconi in 1952. Williams’ et al. described the association of supravalvular aortic stenosis with typical facial features and mental retardation in 1961; this was later supported by Beuren et al in 1962. Between 28 to 50% of supravalvular aortic stenosis have Williams’ Syndrome, which is characterized by Elfin facies, mental retardation, infantile hypercalcemia, short stature and cardiovascular abnormalities.

Recently the genetic basis has been worked out, submicroscopic deletion of chromosome 7q 11.23 in the region of the elastin gene has been found to produce Williams’ Syndrome. This deletion is observed in 90 – 95 % of cases. The mode of inheritance is autosomal dominant. Translocation has also been reported.

The characteristic facies of Williams’ Syndrome is described as “Elfin” facies. The facial features are hypoplastic mandible, small chin, large mouth with patulous lips, blunt upturned

Fig 1. The typical facies of William’s syndrome with small chin, short philtrum, blunt nose, depressed nasal bridge, medial epicanthal fold, hypertelorism with dental malformation and malocclusion

Fig 2. The Image ‘A’ on the left shows the hourglass type supravalvular aortic stenosis during left ventriculogram. The Image ‘B’ on the right shows the stenosis of the right pulmonary artery origin during main pulmonary artery angiogram.
nose, depressed nasal bridge, widely set eyes with internal strabismus, broad forehead, baggy cheeks and dental malformations with malocclusions. These people have a friendly temperament and a deep metallic voice. The skeletal anomalies are an incurving 5th finger, hallux valgus and a long neck.5-7

The children also manifest mental retardation, attention deficit, hyperacusis and hyperventilation. Many children with Williams’ Syndrome seem to have fairly good long-term memory. Young children may be excessively fearful of strangers and especially fearful of being touched.6, 7

The abnormal calcium metabolism seen in a third of these patients is poorly understood. Impaired calcitonin secretion has been suggested as a mechanism in some studies. The hypercalcemia is present in the first year of life and ionized calcium values are more likely to be deviated than total calcium values. Children are prone for rickets as they grow older.8 Renal and renovascular defects have also been described, the defects including renal artery stenosis, segmental scarring, cystic dysplasia, nephrocalcinosis and renal insufficiency.9

The BP difference between both upper limbs in our case is a typical feature of supravalvular stenosis. The selective streaming of blood to the right arm, called Coanda effect, results in better pulse volume and higher BP compared to the left arm and lower extremities.

Cardiovascular abnormalities have been noted in 75 % of the cases, which include supravalvar aortic stenosis, seen in over 50% of the cases. Peripheral pulmonary artery stenosis, valvular pulmonary stenosis, VSD, ASD and atrioventricular septal defects are the other cardiac anomalies described. In 1975, Jones and colleagues reported 19 patients with Williams’ syndrome where five had pulmonary stenosis, two had ventricular septal defect, two had atrial septal defect, and one had valvular aortic stenosis.3 Extra cardiac defects detected were three instances of peripheral pulmonary stenosis, three of aortic hypoplasia, and two of mesenteric and coeliac artery stenosis. Mitral valve disease rarely accompanied Williams’ syndrome. Becker and colleagues reported mitral valve thickening in three cases of supravalvular aortic stenosis.10 The coronary arteries arise before the supravalvular obstruction, which usually starts at the sinotubular junction. Thus the coronaries are exposed to the very high prestenotic pressures leading to accelerated atherosclerotic obstruction and aneurismal deformation. Systemic hypertension, long segment narrowing of aorta with or without coarctation and coronary artery disease leading to myocardial infarction have also been recognised.11

Conclusion

The Williams’ syndrome is a clinical diagnosis that can manifest the typical phenotype and cardiac anomalies, yet present without any cardiac decompensation. This diagnosis requires thorough evaluation of the cardiovascular system. Management of cardiac lesions is predominantly surgical. Association of other cardiac defects needs careful evaluation, usually including catheterization. Outcomes are poorer when more lesions are present. Optimal results depend on appropriate selection and timing of procedures.

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**NEWS AND NOTES**

**CONTEMPORARY ISSUES IN PEDIATRICS**

*January 31 and February 1, 2004*

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