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Education for all is a watchword and a catch phrase, but has a short history of about 150 years. Furthermore it is in serious trouble worldwide. The basic objectives of education are threefold: to prepare people for an increasingly complicated everyday life, to inoculate cultural values and to prepare people for employment in the community. The last objective is usually carried out after schooling through technical colleges and universities. A technical college informs students about a fairly narrow field, whereas a university is far broader in approach, teaching people how to think in addition to informing them. This distinction is often blurred in the world, which favours specialization in a narrow area as a way of dealing with increasing knowledge requirements.

This worldwide educational system has gone awry. Over 20% of young adults in the USA and UK are functionally illiterate, many of them with high school diplomas. A similar proportion in developed Switzerland cannot follow the instructions on an Aspirin bottle. In the developing world the situation is disastrous. Official literacy figures, using inappropriate criteria, overestimate the ability to handle everyday affairs. Educational systems are focussed on note learning and memory. The implications of eliminating teaching people how to think, has greater effects than is generally recognized. At the everyday level, people crossing roads never think of overtaking cars or vehicles approaching from the opposite direction, street sweepers diligently clean steps without thinking of garbage disposal to avoid inevitable reaccumulation; rigidity of training and inability to think makes tradesmen capable of a limited range of tasks. At the national level, the failure to teach people how to think was perhaps the major hidden factor in precipitating the S.E. Asians financial crash. Thailand, wishing to expand and compete with the Asian tigers in computer manufacture, spent about US$ 153 million in a technical skills training programme. Only a few mechanics were produced, so the "hot" money of international investment was withdrawn and the domino effect began. This demonstrates the importance and folly of technical skills training in the absence of an education base. It has little to do with intelligence, as emigrants to the developed world are remarkably successful.

The only Pakistani noble prize winner pointed out that he had to go to the USA to learn how to think; so this is the fundamental defect in developing countries. It affects medicine in two district areas: firstly, the lack of knowledge of the general public is at least partly an ethical responsibility of the medical profession as a whole and doctors individually. Secondly, medical training at the note level produces an intellectual underclass that cannot compete with the developed world in the expanding field of medicine and is an insult to students. Teaching students to diagnose a sore throat and treat it with antibiotics is simply inadequate training. Fast move is demanded of doctors by the international globalized community. They must be aware of patient's circumstances, the underlying causes of recurrence such as nutrition, non-potable water or poor sewerage as well as long term issues such as antibiotic resistance. All of these factors interact in individual patient care. Moreover disease process must be understood in terms of molecular medicine whatever the medical facilities, in order to understand the inevitable conceptual and practical changes in the future. This is the responsibility of the educational system.

In Pakistan, significant efforts are being made to modify teaching methods and ensure uniformity and fairness of the examination process. These efforts deal with the mechanics of medical education and pay lip service to the only essential factor for
improvement of educational outcome viz. that students at university be taught how to think, in addition to being taught how to remember. Reliance on rote learning is entrenched by the school and examination system, which rewards factual recall with high marks. Because of the entrance criteria, all medical students have excellent memories and great potential. The majority simply continue the rote learning system that worked for them at school. Rote learning is, however, merely the first stage in acquiring knowledge and the ability to think is a far more crucial skill in the present day world where a vast and rapidly increasing amount of knowledge needs to be accrued in order to become an internationally acceptable doctor by the WHO criteria. It should be remembered that doctors in Pakistan need to be better than their colleagues elsewhere because of limited facilities and high disease load. The worldwide truism that medical students these days become good doctors despite their training not because of it, is unacceptable in Pakistan – our medical students have to be better doctors than the present generation, because of the future that they will have to deal with.

This can only be accomplished by means of deep learning. Recent advances in the understanding of brain function, largely by use of functional MRI and position emission tomography show that memory and conceptual understanding occur in different areas of the brain and that brain cells do regenerate indeed, like physical muscles exercises, if you do not use it you lose it. The practical application shows very strongly that far more organized information can be recalled by deep learning compared to rote learning. Deep learning is therefore far more efficient and utilizes self testing prior to passive reading and memorization. The emphasis is on understanding starting from a generalized framework rather than the continuation of school day attitudes of memorization of isolated and rapidly changing "facts" from outdated textbooks or formal lectures. Examinations are viewed as a necessary irritant on the way to a life time continuing acquisition of knowledge, rather than being the prime objective, which has the obvious consequences of certification focus, the examination and tutorial industry and widespread cheating. The worldwide trend towards in-course assessment, problem based learning and decrease of formal lectures clearly downrate the examination industry and favour deep learning. Textbooks furthermore, rapidly become obsolete and are best viewed as a framework for knowledge rather than as a source of "facts".

Therefore the introduction of deep learning rather than rote learning is the greatest priority in university education, rather than simply informing students, which is the role of simpler technical colleges. Much resistance to change is encountered among teaching staff, institutions and the students themselves but the nettle must be grasped if deterioration is to be avoided in medical education.

The long-term solution is clearly by teaching children how to think at school. This is an enormous task, which will involve retraining of teachers rather than merely uprating the methodology of teaching, but really dealing with fundamentals in a practical manner rather than reorganization or empty words.

As an interim measure, bridging courses to encourage deep learning are essential on entrance to universities to bridge the gap between school and a true university, of which there are few in the world. As far as medical postgraduate degrees are concerned, certification requirements should include philosophy, computers, genetics, molecular history, biostatistics and many other disciplines prior to award of such degrees to ensure a truly educated person worthy of a university degree rather than a technical diplomat. There is simply no other alternative.

PETER BAILLIE
PREVALENCE OF BACTERIAL VAGINOSIS IN ASYMPOTOMATIC PREGNANT WOMEN AND ITS EFFECT ON THE OUTCOME OF PREGNANCY

RAZIA KOREJO, AHMAR ALI SHAH, KHURSHEED NOORANI

ABSTRACT:
A study was conducted to investigate the prevalence of bacterial vaginosis (BV) in asymptomatic pregnant women attending the ante-natal clinic at JPMC, and to assess the effect of bacterial vaginosis on the outcome of pregnancy, that is pre-term labour, pre-mature rupture of membranes and pre-term birth. Total number of patients in this study was 300, (from March to October 1996). Prevalence of bacterial vaginosis during pregnancy was 20% and pre-term labour occurred in 50% (p<0.001), pre-mature rupture of membranes in 30% (p<0.001) and pre-term births 50% (p<0.001) of these women.

KEY WORDS: Bacterial vaginosis, pre-term labour, pre-mature rupture of membranes

INTRODUCTION
B.V. is a condition characterized by abnormal growth of both anaerobic and aerobic bacteria Lactobacillus, which is dominant in vaginal discharge, is replaced by the over growth of endogenous organisms such as mycoplasma hominis mobiluncus and anaerobic gram-negative rods and gran-positive cocci. Patients often present with thin whitish or greyish-white discharge, which produces discomfort but no itching or pain. Discharge has distinctive fishy odour, it stains underclothes and sometimes causes vaginal bleeding or spotting.

B.V. is not characterized by an inflammatory response. The reported incidence of BV varies in different populations and the range lies between 15-29% in pregnant women. According to Thomson, bacterial vaginosis is diagnosed in 30-35% of cases, making it the most commonly diagnosed condition in the United States, in women who complain of vaginal discharge.

The diagnostic criteria of BV are whitish or greyish-white vaginal discharge which adheres to, but can be easily wiped of from the vaginal wall, pH of vaginal secretions > 4.5, amine test or Whiff test positive and clue cells presence. The presence of any three of the above mentioned four signs is considered to be diagnostic of bacterial vaginosis. Several studies have found a close relationship between BV and premature rupture of membranes, pre-term labour, chorioamnionitis and post partum endometritis. Pre-term onset of labour may be related to large quantities of Phospholipase A2 produced in patients with bacterial vaginosis. A vaginal of pH>4.4 by itself strongly predicts premature rupture of membranes.

PATIENTS AND METHODS
This study was conducted at the antenatal clinic of JPMC from March to October 1996. JPMC is a large hospital at Karachi, which is mainly attended by the under privileged people of the city and where annual patients’ attendance at the Gynaecology & Obstetrics department is 53,000 to 60,000. Patients, who were 18-26 weeks pregnant, attending antenatal clinic for routine check-ups, were included in the study during the said period. Those with a history of abnormal vaginal discharge or itching were excluded. Medical history, clinical examination and obstetrical history were recorded. Patient’s expected dates of delivery was calculated from her last menstrual period and first ultrasound report. Sims speculum was used and vagina was examined to see the colour, amount of discharge and pH

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Prevalence of bacterial vaginosis in asymptomatic pregnant women and its effect on the outcome of pregnancy

of vaginal discharge (with pH paper). Amine test was carried out by adding 10% KOH to the vaginal discharge collected on a slide; a fishy smell indicated a positive amine test. A wet smear was prepared for motile trichromonades, clue cells and yeast. Another vaginal smear was also collected for gram stain and clue cells.

Records of the 300 women included in this study were analysed and results compiled. All patients continued routine antenatal care till their deliveries. Complications of deliveries such as pre-term labour, premature rupture of membranes and pre-term births were also recorded and results compiled. No treatment was given for asymptomatic bacterial vaginosis.

RESULTS
A total of 300 women registered in this study, 60 of them had BV (20%). All these patients delivered between August and October 1996 during which period the total number of deliveries was 1396. The ages of patients ranged from 16 years to 35 years (average 22 years) and parity from 0-8 (average 3). They all belonged to low socioeconomic class and 90% of them were uneducated. None of them were smokers and all had single partners. They outcome of pregnancy is given in Table I

<table>
<thead>
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<th>TABLE-I</th>
<th>OUTCOME OF PREGNANCY IN PATIENTS WITH BACTERIAL VAGINOSIS AND CONTROL GROUP</th>
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<tr>
<td></td>
<td>Control</td>
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<td>---------</td>
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</tr>
<tr>
<td>PTL</td>
<td>n=1396</td>
</tr>
<tr>
<td>PROM</td>
<td>7.8%</td>
</tr>
<tr>
<td>PTD</td>
<td>8.5%</td>
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There was considerable overlap within the patients who had PROM, PTL and PTD. This study indicated that the risk of PTL, PROM and PTD each was approximately 6 times higher than the rest of the population.

DISCUSSION
In the obstetric population of United States, the reported incidence of BV is 10-32%. In Britain the reported prevalence of BV in women undergoing termination of pregnancy is 28%. In this study of asymptomatic pregnant women attending antenatal clinic, BV was found in 20%. The higher incidence of BV in the U.K. and U.S.A. could be due to multiple sexual partners, smoking, pelvic inflammatory disease and urinary tract infections. In JPMC all the patients were non-smokers, with single sex partners who belonged to low social class, with lack of cleanliness. Gardnerella vaginalis and anaerobes associated with bacterial vaginosis are frequent isolates from amniotic fluid and from the amnion and chorion. In fact, bacterial vaginosis is associated with PTL and chorioamnionitis. Although bacterial vaginosis is not considered to be a serious disease or infection threatening the health of a woman, it can and should be prevented. Women can be asymptomatic, or present with symptoms, the unpleasant discharge commonly lead them to seek advice. Individual lifestyle of the patient can strongly determine whether they seek medical attention and this may be highlighted by the fact that women belonging to low social class may not be concerned about the smell or discharge, but may present with itching. The presence of BV during pregnancy indicates the presence of pathogens in the lower genital tract, which are harmful during pregnancy especially when patient's resistance is low. Many studies have shown that patients with BV have a 2.6 times higher risk of going into pre-term labour, 3 times higher risk of premature rupture of membranes and 6.9 times higher risk of pre-term deliveries. In the current study, the risk of PTL, PROM and PTD was approximately 6 times higher than the rest of the population. Screening of all women attending the antenatal clinic is advised to help reduce this risk. Where screening is not implemented, patients suffering from BV should be treated actively.

REFERENCES


A COMPARATIVE STUDY OF LAPAROSCOPIC AND OPEN MINI CHOLECYSTECTOMY

A. SAMAD KHAN, WAQAR ALAM JAN

ABSTRACT:
A comparative study of 160 cases undergoing mini-cholecystectomy without drain (n=80) and laparoscopic cholecystectomy (n=80) was done in Surgical C Unit, LRH from February 1996 to February 2000. All the patients were suffering from calculus and acalculus cholecystitis without complication and were operated on elective basis. Average operation time in laparoscopic cholecystectomy was 1 to one half hour and that of open was 35-45 min. The overall conversion rate was 2.5% in laparoscopic and 1.25% for mini-cholecystectomy group. Average hospital stay was 2 days in laparoscopic cholecystectomy and 4 days with mini-cholecystectomy. Patients who had laparoscopic cholecystectomy returned to normal activity after 10 days whereas patients who had mini-cholecystectomy returned to normal activity after 15 days, on an average.

KEY WORDS: Cholecystectomy, laparoscopic cholecystectomy.

INTRODUCTION
The history of biliary tree stones goes back to pre-Biblical times i.e. to 2000 BC. It was discovered from mummy of a priesten of Amen of 21st Egyptian dynasty and a Babylenian described it (1085-945 BC). Laparoscopic cholecystectomy is being performed in most of the teaching hospitals of Pakistan today and it was started at Lady Reading Hospital four years back. This study was conducted to describe our experience of this modality of treatment.

PATIENTS AND METHODS
All the patients were admitted through OPD, after necessary investigations. A comparative study of cholecystectomy was performed on 160 patients, 80 of which were done laparoscopically and 80 through mini-cholecystectomy incision without drain. This study was conducted between February 1996 to February 2000. All the patients were female with symptoms of gall bladder disease.

Those with mass in right hypochondrium, gall bladder mass or those with common bile duct stones were not included in the study. Detailed history was taken and patients were clinically examined which was documented along with results of investigations and details of operation and subsequent complications. All patients were given prophylactic antibiotics.

RESULTS
Out of 80 patients undergoing laparoscopic cholecystectomy, 75 had gall stones and 5 were having acalculus cholecystitis. In the other group, 60 patients who were operated by mini-cholecystectomy had gall stones and 20 were with acalculus cholecystitis. Maximum number of patients operated by laparoscopic cholecystectomy were 40 in the age group of 40-50 years. The maximum number of patients operated by mini-cholecystectomy were 30 in the age group of 40-50 years. One patient from laparoscopic cholelcyctectomy group was admitted for hepatic artery injury for which we had done conversion to open cholecystectomy. Another patient from the laparoscopic group had sustained a major bile duct injury requiring conversion.

One patient from laparoscopic cholecystectomy group developed collection, which was opened on 3rd post...
operative day. The only patient who was reopened, in mini-cholesystectomy group, developed local abscess which was operated on 8th post operative day. It was observed that the patients undergoing laparoscopic choleystectomy required less analgesia as compared to mini-cholecystectomy in the post operative period. One patient with laparoscopic cholecystectomy developed incisional umbilical hemia but there was no incisional hemia from the other group. Paralytic ileus was seen in 3 patients with laparoscopic cholecystectomy and one with minicholecystectomy. All the patients responded well to conservative management.

Wound infection was seen in 4 patients with mini-cholecystectomy which responded well to conservative management. Average hospital stay was less in laparoscopic cholecystectomy (2 days) while in mini-cholecystectomy average stay was more than 4 days. Patient returned to normal activity in 10 days who had laparoscopic cholecystectomy whereas patients who had mini-cholecystectomy returned to normal activity after 15 days.

DISCUSSION

Management of biliary tract disease has been changed completely as a result of minimal invasive treatment and throughout the world laparoscopic cholecystectomy is a major improvement over open cholecystectomy. In poor countries like ours it cannot be offered to the general public for economical reasons. Mini-cholecystectomy which is a comparable procedure for treatment of gall stone disease can be considered as an alternative to laparoscopic cholecystectomy in the third world. In our study all patients were operated as elective procedure which is safer.

Bile duct injuries are more common in laparoscopic cholecystectomy than in open cholecystectomy. In open cholecystectomy even when abnormal anatomy is encountered one can avoid the risk of bile duct injury by fundus first method. In our study we have encountered one CBD injury in laparoscopic cholecystectomy group. Average time for return to work was 10 days for laparoscopic cholecystectomy and 15 days for mini-cholecystectomy. In our initial practice conversion to open cholecystectomy is 2/80 patients as compared to 1.2-7.5% in some series Some people favour incision cholecystectomy to laparoscopic cholecystectomy as the latter requires expensive equipment and special training.

In our experience if good setup is available for laparoscopic procedure and the surgeon is also experienced, cholecystectomy can be offered to financially sound patients. But in our series, perhaps due to the fact that we were in the learning curve of laparoscopic surgery, the results of mini-cholecystectomies are better than the laparoscopic procedures.

REFERENCES
CORONARY ARTERY BYPASS GRAFTING THROUGH LOWER HALF MEDIAN STERNOTOMY: MODIFIED LSD HOSPITAL TECHNIQUE

ARIF-UR-REHMAN KHAN, SOHAIL BANGASH, MOHAMMED MUSHARAF, M. REHMAN

ABSTRACT:
A study of 42 patients with who underwent coronary artery bypass grafting through lower half median sternotomy, was conducted at National Institute of Cardiovascular Diseases Karachi. The technique that we adopted was a modification of LSD Hospital technique. This report describes detail of this procedure.

KEY WORDS: Technique, Median sternotomy.

INTRODUCTION
Cardiac operations are now being performed through alternative incisions other than standard median sternotomy. Different partial sternotomy approaches have been suggested by various authors. In April 1998 we devised a modification of lower Half T-sternotomy initially evolved at LSD Hospital, Salt Lake City U.S.A. This approach provides adequate exposure of heart for coronary revascularization without any expensive newer equipment or elaborate set-up. It also overcome some of the limitations of original L.S.D. Hospital technique. The present study reports our initial results at National Institute of Cardiovascular diseases, Karachi.

PATIENTS & METHODS
A total of 42 (39 males and 3 females) patients underwent coronary artery bypass grafting using this approach. Age ranged from 34 to 72 years (Median : 57 years). Pre-operative risk factors includes; diabetes mellitus (29%), hypertension (26%), previous myocardial infarction (61%), unstable angina (5%) and COPD(5%).

Angiography revealed left main coronary obstruction in 4 patients, LAD system involvement in 41 patients, circumflex system involvement in 26 patients and diseased RCA system in 38 patients. Ventriculogram showed adequate LV function in 6 patients. Eight patients had mild dysfunction and 2 had severe LV dysfunction.

Technique
A vertical skin incision is made, starting 2 cm below sternomanubrial junction, extending to xiphoid. The pectoralis and intercostal muscle fibers are separated over second intercostal spaces on both the sides. The sternum is divided along an arrow shaped line, with tip of the arrow extending partially into the manubrium and the long arm of the arrow dividing lower sternum in the midline (Figure :1). A medium sized thoracotomy retractor is used for sternal retraction. Internal mammary artery can easily be harvested using the same reactor, by lifting the left sternal border upwards. Dissection of the proximal portion of the artery may require an additional manual retraction of the intact septum by an assistant in some cases. Thymic fat is usually displaced upwards and does not require division of the lobes. Pericardium is vertically opened starting 2 cm below the aortic reflexion up to the diaphragm. Pericardial stay sutures are placed and fixed in such a way that the whole pericardial sac is retracted downwards. Two additional stay sutures are placed on both the sides of aortic reflexion for better exposure of ascending aorta. For extra-corporeal circulation, aorta is cannulized just below the pericardial reflexion. A two-stage venous cannula for inferior vena cava may be introduced through the lateral wall of the right atrium, if two separate venous cannulae are desired. In this case, the inferior cannula is passed through a separate stab incision, below the xiphoid, to avoid obstruction to view operative field. This stab incision will later be used for one of the drain tubes. Once the patient is on extra-corporeal support, ventilation is stopped. Right
upper pulmonary vein is used for vent cannulation. All such patients are being operated at moderate hypothermia alongwith intermittent aortic cross-clamping and ventricular fibrillation. Ordinary size, right angled aortic clamp is used. Proximal end of the graft is anastomosed to aorta during declamp phase. Internal mammary artery (IMA) to left anterior descending anaestomosis is performed last. All anastomoses are performed under direct vision, using standard instruments. All sutures are manually tied. Extra-long instruments are not required.

RESULTS
An average of 3.1 grafts per patients were anastomosed. IMA was anastomosed to LAD in 36 patients. 34 patients had RCA revascularization, 26 had circumflex system bypassed and all the patients had single or multiple grafts in LAD system. One patient required left ventricular plication, as well. Post-operatively, all the patients were electively ventilated in Intensive Care Unit for 2–14 hours. One patient had injury to right internal mammary artery and required re-exploration for excessive bleeding. Average blood loss in rest of the patients was 340ml in the first 24 hours. Other complications included transient conduction defects in 2 patients, unstable union of the upper segment of sternotomy in one patient and superficial wound infection in 2 patients. There was no mortality.

DISCUSSION
There has been a consensus among the cardiac surgeons that most of the morbidity after coronary artery bypass grafting, is not due to surgery of the target organ itself, but due to trauma to the surrounding structures during surgery. Newer techniques are being evolved to minimize this trauma. Main objective is to reduce post-operative morbidity and enhance recovery. In addition to standard median sternotomy and grafting one beating heart, various other options are being evaluated at different centres. Some of these options are surgery via small thoracotomy incision, video-assisted direct surgery and more fancy systems like port-access.

Today, we have two extremes; first, minimally invasive direct surgery on beating heart without any immobilizing mechanism, and second, Ports-Access technique. The former may be a compromise on the precision of anastomosis, with local immobilizing mechanism like “Octopus”, this limitation has been overcome. Still, some of the distal branches of circumflex coronary may not be accessible at the same time. Port-Access technique provides extra-corporeal circulation, cardiac arrest at hypothermia, blood-less operative field and good myocardial preservation. Pre-requisites and limitations of this system are video-assisted thoracoscopy, per-operative fluoroscopy, femora-femoral cannulation, unfamiliar exposure, long operative time and long learning curve.

Between these two extremes, we may put lower half sternotomy approach. The procedure proposed originally by workers at LSD Hospital was a T-shaped sternotomy, with horizontal limb of ‘T’ lying just below sternomanubrial junction. In our experience, this sternotomy had three drawbacks:

- Lifting of upper part of sternum required separate retraction with Favaloro IMA retractor,
- A separate incision was needed for passing aortic clamp,
- Post-operatively upper end of the sternotomy remained unstable because of the slide-ways movement between upper and lower halves of sternum in many cases.

To overcome these problems, we hypothesized that converting the horizontal limb of ‘T’ into an arrow-head whose apex should extend into manubrium, we could make the procedure more simple and less cumbersome. Post-operatively, the lower two halves could jointly wedge into the upper intact sternum. Better visibility of upper part of ascending aorta and extra space for application of aortic cross clamp without obstructing the view are additional advantages. The approach itself provides a small incision with a familiar operative exposure. One small to medium wound replaces multiple small wounds in chest, groin, and neck. Ordinary paediatric size internal defibrillator paddles are used. It can easily be converted into standard median sternotomy in case of emergency. Complete revascularization can be achieved using all traditional grafts like both IMAs, saphenous vein and radial artery. No special or extra-long instruments are required. There is no difficult learning curve involved. Post-operatively, intact upper sternum, weight bearing area is much stable, with minimum possibility of brachial plexus injury. Post-operative bleeding is remarkably less. Low midline scar is cosmetically appreciated by the patient.

There are however, a few limitations of this techniques. Surgery is more tedious and slow due to small size incision. Achieving operating rhythm requires more experience. Surgery is more difficult in obese or deep chested patients and those with diluted or hypertrophied ventricles. In conclusion, this is an excellent alternative to routinely used median sternotomy, which can be learnt easily. So far our results have been encouraging.

REFERENCES


ABSTRACT:
Recurrent ventral hernia is not an uncommon problem. Most of the patients present with discomfort and unacceptable appearance. A few develop risk of strangulation or obstruction. Repair in these cases is challenging with increased risk of respiratory failure, infection, enterocutaneous fistulae and above all recurrence. In this study of 17 patients with recurrent incisional hernia, extraperitoneal, sub-muscular mesh repair was done with no recurrence. Complications like wound infection (31.2%), respiratory failure (12.5%), paralytic ileus (68.7%), seroma (6.2%) and sinus (31.2%) were encountered.

KEY WORDS: Recurrent ventral hernia

INTRODUCTION
Recurrent ventral hernia is always challenging for surgeons due to its peculiar symptoms, etiological factors, abdominal wall defects, dense bowel adhesions, previous surgical failures and impending risk of recurrence. (Figure 1)

Specifically, a recurrent ventral hernia is one that develops at the site of previously repaired hernia in the anterolateral abdominal wall.1

Studies on incisional hernia repair have shown recurrence rate of 20-40% using apposition technique in comparison to less than 10% when mesh repair is done.2 The other techniques in use are Keel's operation, Fascia lata repair, Tantalum repair and Synthetic (nylon, polyester or polypropylene) repair are becoming more popular in practice, because of its easy availability, cost effectiveness and low recurrence rate.3

The mesh, being a foreign material, may increase the risk of infection, adhesions, sinus formation, enteric fistulae and extrusion of mesh. Prosthetic repair was introduced in 20th century and Uscher et al were the first to introduce repair by Marlex mesh.4

PATIENTS AND METHODS
This prospective study of mesh repair was started on 1st June 1997 and continued up to 30th December 1999. All adult patients of either sex, suffering from recurrent ventral hernia, admitted in the Department of Surgery, Bahawal Victoria Hospital, Bahawalpur were included in the study. Patients were selected on the basis of failure of primary or secondary repair. A proforma regarding epidemiological data, previous date and type of surgery alongwith possible recurrence time, physical signs of the hernia, date and site of prosthetic implant and recording for 4 months was completed.
RESULTS
A total of 17 patients were included in the study. One patient could not be offered repair even with recurrent attacks of obstruction due to chronic obstructive pulmonary disease (COPD). In this patient obstruction was relieved by enlarging the defect and no repair was done. Out of 16 patients, 10 were female and 6 male. Average age was 53 years (Range 40-70 years). In all the patients previous repair was done with prolene, except one patient where prolene mesh was applied subcutaneously over rectus sheath. In most of the patients repair had been attempted once, while in 2 patients it had been attempted twice (Table I)

<table>
<thead>
<tr>
<th>Indication for Surgery.</th>
<th>No. of Patients</th>
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<tr>
<td>Discomfort</td>
<td>17</td>
</tr>
<tr>
<td>Enlargement</td>
<td>17</td>
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<tr>
<td>Unacceptable</td>
<td></td>
</tr>
<tr>
<td>Appearance</td>
<td>17</td>
</tr>
<tr>
<td>Risk of strangulation</td>
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<tr>
<td>Strangulation/Obstruction</td>
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</tr>
<tr>
<td>Skin Conditions.</td>
<td></td>
</tr>
<tr>
<td>Normal</td>
<td>5</td>
</tr>
<tr>
<td>Infection/ulceration</td>
<td>11</td>
</tr>
<tr>
<td>Enterocutaneous fistula</td>
<td></td>
</tr>
<tr>
<td>Size of Defect</td>
<td></td>
</tr>
<tr>
<td>Less than 6cm.</td>
<td>Nil</td>
</tr>
<tr>
<td>6-10cm.</td>
<td>12</td>
</tr>
<tr>
<td>More than 10cm</td>
<td>5</td>
</tr>
<tr>
<td>Reducibility</td>
<td></td>
</tr>
<tr>
<td>Reducible</td>
<td>Nil</td>
</tr>
<tr>
<td>Irreducible</td>
<td>17</td>
</tr>
<tr>
<td>Mesh Placement.</td>
<td></td>
</tr>
<tr>
<td>Without anterior rectus</td>
<td>11</td>
</tr>
<tr>
<td>With anterior rectus</td>
<td></td>
</tr>
<tr>
<td>sheath closure</td>
<td>5</td>
</tr>
<tr>
<td>Drain (Removal)</td>
<td></td>
</tr>
<tr>
<td>Within one week.</td>
<td>8</td>
</tr>
<tr>
<td>Upto 14 days or more.</td>
<td>8</td>
</tr>
<tr>
<td>Antibiotics.</td>
<td></td>
</tr>
<tr>
<td>For one week.</td>
<td>5</td>
</tr>
<tr>
<td>Two weeks</td>
<td>6</td>
</tr>
<tr>
<td>Three week or more</td>
<td>6</td>
</tr>
</tbody>
</table>

In all the patients two redivac drains, one under mesh and another in the subcutaneous space, were placed. Respiratory failure was observed in 2 patients which was managed by prolonged active nasogastric aspiration, oxygen inhalation and other respiratory supportive measures. Paralytic ileus was seen in 11 patients. Seroma occurred in one patient and severe wound infection was seen in 5 patients, who were successfully treated by antibiotics according to culture sensitivity reports. Sinus occurred in 5 patients and on the lateral side of implant within 8-15 weeks of repair which were treated by removing the underlying prolene stitch and antibiotics, but in no case mesh was removed or its extrusion occurred. No recurrence was seen.

DISCUSSION
Recurrence rate of incisional hernia repair may be as high as 20%6,7,8 and 80% of these recur within 2 years. Incisional hernia is a relatively middle age female problem (50-60 years)2,6,7,14 as was confirmed by this study. Polypropylene mesh has been the most widely used prosthetic material in repair of hernia since it was first introduced in 1963,7,8,10 But it can cause various complications due to its physical properties i.e. adhesions, enteric fistula, erosion into intra-abdominal organs and mesh extrusion etc. To minimize all these complications, we placed the mesh in extraperitoneal space under the muscles.10 For tension free closure, peritoneum had been dissected from the hernial sac and anterior rectus sheath had not been closed in all cases, rather it was stitched to the mesh (Figure 2).

In comparison to polypropylene mesh, PTFE (polytetrafluoroethylene) mesh is nonabrasive, minimally irritant (inert) with few chances of bowel/organ adhesions or enterocutaneous fistulae and are strong enough to support. Thus there will be remarkable reduction in postoperative complications.6,12 But we cannot use PTFE mesh due to its limited availability in Pakistan. Moreover experience with PTFE is very limited and short, so long term follow-up data has yet to become available.6,12,13,14

The most common postoperative complication was paralytic ileus (68.7%) which is very high in comparison to literature. It may be due to extended adhesiolysis and abdominal wall dissection. This is said to be common
cause of recurrence, but no recurrence was noted in this study.

Post operative wound infection was found in 5 (31.2%) cases which is again higher. It may be due to repeated surgeries, large dead space and preoperative skin ulceration and infections (68.7%). Better preoperative wound care, proper use of antibiotics, avoidance of dead space by Paseal's principle and use of suction drains are the mainstays of prophylaxis. This high infection rate may be due to missed seroma under the muscles. All these cases were treated by wound debridement, wound dressings and antibiotics according to culture sensitivity reports.

Seroma formation was noted in one (6.2%) case, that is less than standard i.e. 14.5%. It should be aspirated by aseptic technique. Sinus formation noted after 8 to 15 weeks of repair in 5 (31.2%) cases. This is also high incidence, probably due to missed sub muscular seroma or foreign body stitch reaction. Respiratory failure was the most dangerous and difficult to manage complication. It was observed in 2 (12.5%) cases in repair of very large hernias with loss of right of domain. Such cases should be properly prepared preoperatively. But despite all efforts these cases need post-operative ICU facilities and positive pressure ventilation. Preoperative pneumoperitoneum to increase peritoneal cavity is obsolete.

**CONCLUSION**

The repair of large incisional hernias and recurrent incisional hernias with synthetic sutures or patch is possible beyond any doubt but inability to close the fascia without excessive tension or unrealistic repair due to insufficient tissue, infection or haematoma will lead to recurrence. The recommended synthetic material depends upon the personal choice and availability. We recommend repair of all recurrent incisional hernias with synthetic patch irrespective of the size of the defect to avoid the risk of recurrence. It should be implanted extra-peritoneally under the muscles to avoid adhesions, extrusion and abdominal protrubance.

To avoid infection, meticulous wound care, wound dressings and patient shower twice a day with germicidal/fungicidal solutions, is recommended. Prophylactic antibiotics should be used till the time of removal of drains. Suction drainage, early recognition of seroma/haematoma with aseptic drainage or aspiration, all help to reduce the incidence of infection. To avoid delayed sinus formation, antibiotics incorporated biomaterials and relatively inert material (PTFE) should be used. Good anaesthesia support during and in the immediate post-operative period with ICU facilities is the mainstay of treatment to avoid deaths due to respiratory failure.

**REFERENCES**

PERCUTANEOUS NEPHROLITHOTOMY

ABDUL RASHEED SHAIKH

ABSTRACT:
Over a period of 7 years from 1989 to 1996, 100 patients (70 male and 30 female) had PCNL treatment at Pakistan Institute of Medical Science (PIMS) Islamabad. Their mean age was 32±6.3 years (range 18-55 years). Patients with single or multiple renal calculi measuring less than 2 cm, preferably located in the renal pelvis, were selected for the study. Success rate was 94%. Operative and postoperative complication occurred in 12% and 18% of the cases respectively. Hospital stay was significantly lower and treatment was cost effective. Long term followup did not reveal any problem attributable to the procedure. It is concluded that percutaneous nephrolithotomy is an effective, safe and cost effective procedure and still has a place in the era of ESWL (Extra-Corporeal shock wave lithotripsy).

KEY WORDS: Renal stone, Percutaneous nephrolithotomy

INTRODUCTION
Nephrolithiasis is a common surgical problem. Surgical therapy of symptomatic renal calculi has changed dramatically over the last two decades. The decision for treating a patient with stone is not yet codified and it is configured according to the types of equipment available, size of stone, needs of the patient and skill of the surgeon.1

PCNL allows direct access to the kidney for removal of stone via purpose-made nephrocutaneous tract. The method of Percutaneous nephrostomy was described by Goodwin and associates in 1955,2 but it was not until 1976 that the first PCNL was reported.3 Later this technique evolved and flourished in the early 1980's when large successful series of PCNL extractions were reported. Since then PCNL has been widely accepted as a useful technique and accessory modality for ESWL.4 The concept of PCNL is still new in Pakistan. Our analysis was aimed to ascertain the efficacy, safety, reliability and cost of effectiveness of PCNL. We also explored the prospects of PCNL in Pakistan and its future use.

PATIENTS AND METHODS
This study was carried out at PIMS Islamabad from Feb: 1989 to Dec: 1996. One hundred selected cases were included in the study. The criteria for selection of patients was presence of single or multiple renal calculi measuring less than 2 cm, located in the renal pelvis.

Patients with staghorn calculi in the upper and middle calyx or with associated renal anomalies were excluded from the study. Screening workup included complete history, clinical examination and investigations to see fitness for operation, determine the stone size, assess renal function and detect urinary infection. The investigation included urine examination, culture sensitivity, complete blood counts, biochemistry, ultrasound and intravenous urography.

Under general anaesthesia the collecting system was opacified through retrograde ureteric catheterization. The kidney was punctured under fluoroscopic control in prone oblique position. The tract was dilated up to 32-36F with graded teflon dilators, passed over guide wire. An Amplatz working sheath was passed over the last dilator, which acted as a conduit between skin and collecting system. Nephroscopy was performed and stone was located. Stones less than 1.5 cm were removed intact, whereas larger stones were disintegrated with electrohydraulic or ultrasonic lithotripter. At the end of the procedure, nephrostomy tube was left in place. This tube was removed when drainage became clear. All the patients were requested to attend outpatient's clinic at one, three and six month's intervals for followup.

RESULTS
There were 70 males and 30 were females. Their ages ranged between 18-55 years (mean 32 ± 6.3 year)
Whitfield observed that PCNL is a time consuming procedure but comparable to experience, which is justified by him as his studies included patients with large and staghorn calculi. It is experience, which will help to attain the expertise.

Success rate was achieved in 94% of patients. Stones were removed intact in 84% cases and 8% patients required in situ lithotripsy with electrohydraulic and ultrasound lithotriptor. In 8 patients stones could not be removed. Of them 2 patients under went a second stage PCNL with success, whereas in the remaining six surgery was advised.

Mean duration of the procedure was 90 minutes (± 8.37), excluding anesthetic induction and recovery time. The operative complications included primary haemorrhage (4%) and extravasation of urograffin (4%) and prolonged nephrostomy drainage were observed in 4% cases. Post-operative complications included delayed haemorrhage in 2% patients. It occurred on the third postoperative day with the removal of nephrostomy tube. It stopped successfully by immediate repositioning of the tube, by its tamponade effect. Fever in 12%, chest infection in 6% and urosepsis in 2% were also noticed. Prolonged ileus and vomiting occurred in another 4% cases. Nephrotoneous fistulae occurred upto 6 days in 2% of patients after removal of the nephrostomy tube. The mean hospital stay was 5.7(± 1.08) days (range 4-10 days) and the mortality rate was zero. 56 patients came for followup and 10 were selected for followup intravenous urography and isotopic study between 3-6 months to see effect of PCNL on outcome of the procedure.

DISCUSSION

In our study the overall success rate was 94% which is comparable to Bapat and White. However, Reddy and Segura reported 99% and 98.3% success rates respectively. The difference could be explained by available expertise and equipment. This was our first experience, which will help to attain the expertise. Whitfield reported 65%, success rate with his early experience, which is justified by him as his studies included patients with large and staghorn calculi. It is observed that PCNL is a time consuming procedure but that is not a justification for neglecting the technique. We believe that, as experience grows, the time taken will certainly be reduced.

Overall operative and post operative complications are similar to Hunter and lower than that reported by Whitfield who showed 20% complication rate. Primary haemorrhage occurred in 4% patients, which was intractable in 2% cases, requiring abandoning of the procedure. In these cases blood transfusion was required. Our result is comparable with Wickham and Bapat. In our series frequency of haemorrhage is higher than Segura and Patterson. This complication can be minimized by suggestions given by Patterson that the number of initial punctures should be kept to the minimum and excessive intrarenal manipulation avoided to prevent haemorrhage.

In our series, overall infection rate was high but was lower than Charton. This could be due to non-usage of prophylactic antibiotic by Charton. We agree with their conclusion that PCNL is associated with a risk of infection, thus prophylactic antibiotic cover during and after the procedure in essential. In our study mean duration of hospital stay was significantly low, which is considerably less than compared to that of Reddy. Their series include a large number of patients with staghorn calculi, who required multiple sessions. It is interesting that Preminger. Reported series of PCNL on outdoor basis with no hospitalization at all. We did not follow this because we are not confident that our instructions will be followed by the patients at home.

The most important benefit of PCNL is that it is cost effective. Although initial cost of installment of equipment is quite high but it costs less to patient and less medication is needed. Less hospital stay undoubtedly reduced the cost and another significant economical factor is early return of patients to their jobs. Patients were more satisfied with the outcome of this procedure.

The value of percutaneous approach is not limited to stone removal but is also useful for various other therapeutic and diagnostic purposes. In the era of ESWL, the PCNL serves as pre requisite, not only to debulk the large stone but also to manage the complication of ESWL, like obstruction at ureteric level caused by fragments. Combined ESWL and PCNL is recommended by many investigators because it almost completely eliminates the need for open surgery and achieves high success rate. The experience of PCNL can also be applied for the drainage of the obstructed kidneys, antegrade ureteric stenting. Recently endopyelotomy (percutaneous pyelolysis) has become a new option as an alternative to pyeloplasty. This is another therapeutic use of percutaneous surgery. There are also future
Percutaneous nephrolithotomy

prospects of percutaneous resection of renal pelvic tumors. Jones performed PCNL in solitary kidney in 53 patients with 77.3% success rate. He advocated that it is also a safe procedure in solitary kidney.

From our result and review of literature, we conclude that PCNL is safe and cost effective surgical procedure with low morbidity. It is less traumatic and creates no cosmetic problem. Its experience can be used for variety of other diagnostic and therapeutic purpose. As the incidence and prevalence of urolithiasis is quite high in our country, we feel justified to say that PCNL should be established in every urological center. This could be more useful for our nation if implemented at a wider scale at district level hospitals in future.

REFERENCES
Efficacy of Cyclosporine in 36 Patients with Rheumatoid Arthritis

M. Ashraf Memon, Anisuddin Bhatti

ABSTRACT:
Rheumatoid arthritis is a chronic inflammatory disease that leads to early crippling if its progression is not halted. A prospective study of 36 patients with rheumatoid arthritis treated with cyclosporine A was carried out in Orthopaedic Department of Jinnah Postgraduate Medical Centre from April 1997 to August 1998. We found improvement in more than 90% of patients and 63% of these patients qualified American College of Rheumatology (ACR) criteria for improvement. Drop-out rate in our study was 27%, 8%, due to inefficacy of drug and 19% due to toxic effects.

KEY WORDS: Rheumatoid arthritis Efficacy, Cyclosporine A

INTRODUCTION
Rheumatoid arthritis is a chronic systemic inflammatory disease chiefly affecting the synovial membranes of multiple joints and leads to substantial disability, loss of productivity and increased mortality. Prevalence of rheumatoid arthritis is approximately 1% (range 0.3-2.1%). Females are three times more frequently affected than males. The onset of disease is most frequent during the fourth and fifth decade of life and 80% of all patients developing the disease are between the age of 35 and 50 years. Susceptibility of rheumatoid arthritis is genetically determined. Most patients have class-2 human leukocyte antigen (HLA) with an identical five amino acid sequence. It is four times more common in first degree relatives of individuals with seropositive disease.

Treatment objectives in rheumatoid arthritis patients are to alleviate symptoms and to curb joint destruction. In recent years cyclosporine A has been increasingly used early in the treatment of rheumatoid arthritis to slow down the destructive process and to preserve the joint function. Use of multiple disease modifying agents (Gold, sulfasalazine, methotrexate and anti malarials) maintained the low level of inflammation but this strategy did not control the development of new erosions; number of erosions and disability increased. Efficacy of cyclosporine on a clinical, laboratory and radiological abnormalities has been well established. Low dose of Cyclosporine A significantly reduced the rate of progression of erosion in both early and long standing rheumatoid arthritis compared with disease modifying agents and provide an effective control of symptoms.

Cyclosporine A inhibits transcription of genes encoding interleukin-2 (IL-2) and other cytokines (IL-1, IL-6, TNF a, and INFg). Cyclosporine A also inhibits other cell types including B-cells, macrophages, fibroblasts and chondrocytes involved in rheumatoid joint destruction. Present study is based on the use of Cyclosporine A in the treatment of rheumatoid arthritis with the aim to evaluate the efficacy, safety, tolerability and prevention of further destruction of joint surfaces and deformities.

PATIENTS AND METHODS
Thirty six patients were registered in the Orthopaedic Department of Jinnah Post Graduate Medical Center, Karachi from April 1997 to August 1998, who fulfilled the revised criteria for rheumatoid arthritis laid down by the American Rheumatism Association now known as American College of Rheumatology, (age less than 65 years, normolensive, normal hepato-renal status, no concomitant disease and patients who had not taken
Efficacy of cyclosporine in 36 patients with rheumatoid arthritis

other DMARD or had intra articular steroid injection one month prior to Cyclosporine A therapy).

Before starting the Cyclosporine A detailed history was taken and patient was thoroughly examined and investigated for hepatic and renal functions. Radiological assessment of hand, foot, ankle and other inflamed joints was carried out. Number of tender and swollen joints was recorded. Global assessment by the physician and patient was noted. Pain (as assessed on visual analogue scale) and degree of disability (as measured by Health Assessment Questionnaire) were observed. Erythrocyte sedimentation rate (ESR), morning stiffness and grip strength were included for assessment.

Patients were called for follow up every two weeks for the first three months and then every month for rest of the study period. All the parameters were recorded and patients were investigated for any toxicity.

Cyclosporine A was started in a dose of 3mg/Kg body weight (BW) / day in two equal divided doses at 12 hourly interval. The dose was tailored with response at 2-4 weeks interval upto a maximum dose of 5mg/Kg BW/day. If the response was good and patient improved then maintenance dose of 2-3mg/Kg BW/day was continued for the rest of study period. Improvement was assessed by criteria laid down by American College of Rheumatology. Improvement must occur in three of the following five criteria. - degree of disability, pain, ESR, patient global assessment, physician’s global assessment and + 20% improvement in the number of swollen and tender joints.

Six patients discontinued treatment before completing three months of Cyclosporine A therapy. Another four patients discontinued treatment before six months. Reasons for discontinuing treatment are given in (Table-I). None of the patients discontinued treatment thereafter. Out of the remaining twenty six patients, 14 patients completed one year treatment and twelve patients completed more than six months of treatment to date.

TABLE-I REASONS FOR DISCONTINUING CYCLOSPORINE A THERAPY

<table>
<thead>
<tr>
<th>Reason for discontinuing</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lack of efficacy</td>
<td>3 (8.1%)</td>
</tr>
<tr>
<td>Renal function impairment</td>
<td>2 (5.4%)</td>
</tr>
<tr>
<td>Hypertension</td>
<td>1 (2.7%)</td>
</tr>
<tr>
<td>Impaired liver functions</td>
<td>1 (2.7%)</td>
</tr>
<tr>
<td>G.I. Intolerance</td>
<td>3 (8.1%)</td>
</tr>
<tr>
<td>TOTAL</td>
<td>10 (27%)</td>
</tr>
</tbody>
</table>

RESULTS

Out of 36 patients, 28 were females and 8 males with mean age of 38.6 years (range 20-58 years). Duration of symptoms ranged from 1.5 years to 18 years (mean 12.5 years). On the basis of functional capacity one patient was Class 1, 32 Class II and 3 Class III. Duration of treatment with Cyclosporine Aranged from 6 months to 16 months with the mean duration of treatment 10.5 months.

Outcome of treatment at six months and one year completion of cyclosporine A therapy is shown in Table-II and is compared to the base line, i.e. before the start of treatment. After six months of treatment there was 32% improvement in tender joint count and 28% improvement in swollen joint count. Improvement as assessed by physician was 20% and patient himself was 22%. Improvement in joint pain was 26%, and in the degree of disability 28%. However, ESR changed little and it reduced 2.1 mm first hour. There was marked improvement in the morning stiffness, which reduced from 75 minutes to 18.5 minutes. Grip strength increased from 104 mm of Hg to 216 mm of Hg. All of these parameters either further improved or remained stable except E.S.R. which increased by 2.4 mm. after completion of 12 months therapy (Table-II).

DISCUSSION

There is no drug which can cure rheumatoid arthritis. Optimal management requires early diagnosis and appropriate treatment with DMARDs. Fuchs et al have shown that seropositive rheumatoid patients have a high probability of developing joint damage within two years of the disease onset. Vander HA et al have suggested that early aggressive treatment of rheumatoid arthritis may alter the disease course and prevent progression of joint damage. Many studies have proved the efficacy of immunosuppressive agents like methotrexate and
Cyclosporine in the treatment of rheumatoid arthritis. Methotrexate is an effective treatment for rheumatoid arthritis but some patients response only partially to it.\textsuperscript{10}

As compared to other studies female / male ratio is somewhat higher in our study 3.5:1. Whereas female / male ratio is 2.4:1 in the study of Michael Stein\textsuperscript{10} and Tugwel\textsuperscript{11} and 3:1 in the study of Drosos\textsuperscript{12} Mean age of patients in our study is lower, 39 years as compared to other studies\textsuperscript{10-12} where it is 50 to 55 8years. The difference may be due to the fact that we have not included patients over 65 years of age in our study. We noted substantial improvement in more than 90% of patients treated with Cyclosporine A in our study. These results coincide with the results of Drosos\textsuperscript{12} which have shown similar high rate of efficacy. However only 63% patients qualified the criteria for improvement laid down by the American College of Rheumatology.\textsuperscript{7} Improvement in morning stiffness and grip strength is similar to that observed in the study of Drosos\textsuperscript{12} and Uppulari.\textsuperscript{13} This difference may be due to the short mean duration of the disease in these studies as compared to our patients.

Ten patients (27\%) discontinued treatment. The dropout rate was comparatively low in our study as compared to 46\% dropout rate in the study of Carpenter.\textsuperscript{14} Main cause of discontinuation of treatment in our study was lack of efficacy 30\% and G.I intolerance 30\% followed by renal impairment 20\%, impaired liver function 10\% and hypertension 10\%. Where as the main cause of discontinuation in the study of Carpenter\textsuperscript{14} was renal impairment followed by tremors and hypertension. The difference may relate to the higher mean age group in the study of Carpenter.\textsuperscript{14}

Cyclosporine provides a good option in the treatment of early stage rheumatoid arthritis to slow down destruction of and preserve the joint functions. It has relatively low incidence of side effects specially in the young age group. The efficacy is maintained for prolonged period of time. While most of the side effects occur during the first 6 months of therapy no side effect is added after this period on continuation of therapy.

REFERENCES
HAEMODIALYSIS: ARTERIOVENOUS FISTULA VERSUS INTRAVENOUS CANNULA

MUNAWAR JAMIL, M. SALEEM, S.A. TABASSUM, M ZUBAIR, ASGHAR ALI

ABSTRACT:
A three years prospective study was carried out from July 1996 to June 1999 to compare the results of haemodialysis by arteriovenous (A.V.) fistula versus intravenous cannula (IVC) at Haemodialysis Unit, Department of Surgery, B.V. Hospital, Bahawalpur. During this period a total of 4600 dialysis on 843 patients were performed. 2760 (60%) were done by I.V. and 1840 (40%) by A.V. fistula. Inadvertent arterial puncture in 10 cases (1.9%), cardiac arrhythmia in 5 (0.9%), pneumothorax in 5 (0.9%) and thrombus blockage in 20 (3.9%) were the complications noted on I.V. cannulation. Fever and shivering occured in 75 (15.8%) patients of I.V. cannula as compared to 10 patients (3.2%) with A.V. fistula. Infection at cannula site with I.V. cannula was noted in 50 (10%) and bleeding in 50 (10%) patients. In cases of A.V. fistula aneurysmal dilatation was seen in 10 (2.9%), aneurysm rupture in one (0.29%) and A.V. fistula failure in 8 (2.36%) patients. On an average blood flow in case of AV fistula can be easily maintained up to 300 ml/min. as compared to 250 ml/min. with I.V. cannula. Serum urea and creatinine were more effectively decreased by AV fistula. Mortality at the time of cannulation was 0.4% and none with AV fistula. AV fistula is a far better vascular access for chronic renal failure, as dialysis is more smooth, cost effective and has less complications.

KEY WORDS: Haemodialysis, AV fistula, IV cannula

INTRODUCTION
Acute as well chronic renal failures are common in the surrounding areas of Bahawalpur. End stage renal diseases (ESRD) are on an increase during the past few years.1 Haemodialysis is the most commonly used treatment modality for chronic renal failure in our setup. Expected five years survival for those on haemodialysis is 36%.2 Peripheral AV fistula and IV cannula are methods for providing access for dialysis circulation. According to literature the vascular access in 90% patients on haemodialysis should be maintained by AV fistulas and only 10% patients should be maintained on IV Cannula as permanent chronic dialysis access. Chronic catheter access is defined as use of dialysis catheter for more than 3 months.3

In our centre 60% of the patients are dialysed by IV cannula. Most of the patients of chronic renal failure come irregularly, usually in emergency from the periphery and disappear after dialysis until next emergency situation. They do not comply with the advice for formation of AV fistula. A study was carried out to analyse the results of haemodialysis by AV fistula versus IV cannula.

MATERIAL AND METHODS
All diagnosed cases of renal failure, acute and chronic, are referred to our haemodialysis unit. IV cannula is passed by Sedinger technique in subclavian internal, jugular or femoral vein, keeping in view all the measures of sterilization.

AV fistula is formed at the wrist between radial artery with cephalic vein. Under local anaesthesia, side to side or end to side anastomosis is done with prolene 6/0. AV fistula matures after 6-8 weeks. Bruit indicates that AV fistula is functioning. Haemodialysis is done for 4 hours in all the patients. Dialysis of co-efficient 5-3 is used. Blood is maintained 250 to 300 ml/minute during the dialysis. Heparin 5000 units 1/2cc is injected into the lines at the start of dialysis. Then 1cc heparin diluted in 20cc saline is continuously given during the dialysis. B. Bran dialysis
solution is used in all the patients. Dialysis solution contains K 2.6 mEq/L with high glucose content. Transmembrane pressure (TMP) was adjusted accordingly, fluid removed in one hour equals co-efficient of dialyser multiplied by TMP. Predialysis and postdialysis urea and creatinine were checked to assess the efficacy of haemodialysis.

RESULTS
Eight hundred and forty three patients were dialysed 4600 times over a period of three years. Forty three patients of acute renal failure were dialysed by intra-venous cannula. Eight hundred patients of chronic renal failure, were dialysed either by IV cannula or AV fistula. Out of 4600 dialyses, 2760 (60%) of dialyses on 505 patients were done through IV cannula and 1840 dialyses (40%) on 338 patients through AV fistula.

Efficacy of the haemodialysis by AV fistula versus IV cannula can be compared at vascular access, during dialysis and post dialysis. Complications of IV cannulation or formation of AV fistula, are shown in Table I.

<table>
<thead>
<tr>
<th>Complications at Vascular Access</th>
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<tbody>
<tr>
<td><strong>Complications of IV cannula</strong></td>
</tr>
<tr>
<td>Inadvertent arterial puncture</td>
</tr>
<tr>
<td>Cardiac arrhythmias</td>
</tr>
<tr>
<td>Thrombus formation</td>
</tr>
<tr>
<td>Embolism</td>
</tr>
<tr>
<td>Pneumothorax</td>
</tr>
<tr>
<td><strong>Total</strong></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Complications of AV fistula</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac arrhythmias</td>
</tr>
<tr>
<td>Thrombus formation</td>
</tr>
</tbody>
</table>

The working of both the procedures during dialysis are reflected in Table II.

<table>
<thead>
<tr>
<th>Comparison during Dialysis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>IV cannula</strong></td>
</tr>
<tr>
<td>Blood flow &lt; 250 ml/minute</td>
</tr>
<tr>
<td>Venous pressure &gt;200</td>
</tr>
<tr>
<td>Shivering and fever</td>
</tr>
<tr>
<td>Adverse effect on dialysis by change of position of the patient</td>
</tr>
<tr>
<td><strong>AV fistula</strong></td>
</tr>
<tr>
<td>Blood flow &lt; 250 ml/minute</td>
</tr>
<tr>
<td>Venous pressure &gt;200</td>
</tr>
<tr>
<td>Shivering and fever</td>
</tr>
<tr>
<td>Adverse effect on dialysis by change of position of the patient</td>
</tr>
</tbody>
</table>

Post dialysis complications are compared in Table III.

<table>
<thead>
<tr>
<th>Complications in Post-Dialysis Period</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Complications</strong></td>
</tr>
<tr>
<td>Infection at cannula site</td>
</tr>
<tr>
<td>Aneurysm formation</td>
</tr>
<tr>
<td>Bleeding</td>
</tr>
</tbody>
</table>

Overall differences of Haemodialysis: IV cannula versus AV fistula are compared in Table IV.

<table>
<thead>
<tr>
<th>Overall Comparison</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Factors</strong></td>
</tr>
<tr>
<td>Urea and creatinine</td>
</tr>
<tr>
<td>Decreased 30% to 45% after dialysis</td>
</tr>
<tr>
<td>Decreased 35% to 55% after dialysis</td>
</tr>
<tr>
<td>Cost</td>
</tr>
<tr>
<td>Rs. 2000-2500 for every 4-5 dialysis</td>
</tr>
<tr>
<td>Rs. 1000-1500 for every dialysis</td>
</tr>
<tr>
<td>Time required for the procedure</td>
</tr>
<tr>
<td>15 minutes</td>
</tr>
<tr>
<td>1 hour</td>
</tr>
<tr>
<td>No. of dialysis</td>
</tr>
<tr>
<td>Only 5-6 dialysis with one cannula</td>
</tr>
<tr>
<td>Life long</td>
</tr>
<tr>
<td>Failure</td>
</tr>
<tr>
<td>Nil</td>
</tr>
<tr>
<td>Failure</td>
</tr>
<tr>
<td>2 (0.4%)</td>
</tr>
<tr>
<td>8 out of 338 AV fistulas failed</td>
</tr>
<tr>
<td>Mortality due to procedure</td>
</tr>
<tr>
<td>Nil</td>
</tr>
</tbody>
</table>

DISCUSSION
Unfortunately end stage renal disease is progressively increasing in the Bahawalpur region, same as reported in the western literature. Among the available options for treatment are kidney transplantation and dialysis. Kidney transplantation is the treatment of choice in terms of patient survival and quality of life. But most patients remain on dialysis due to shortage of donors. Among hemodialysis and peritoneal dialysis, the latter depends upon patient's cooperation and education. Our patients usually come from far flung areas and are generally illiterate. It is very difficult to train them for Chronic Ambulatory Peritoneal Dialysis. So in our set up, almost all patients of chronic renal failure are managed on hemodialysis.

The results of hemodialysis are directly dependent on vascular access for dialysis, blood flow rates are sufficient for adequate clearance of uraemic toxins. Vascular access surgery is the most frequently performed procedure for chronic renal failure. Patients should be evaluated before access surgery for proper site. Physical examination, venography, doppler, ultrasound and arteriography, are needed. There are two available options for vascular access: Peripheral AV fistula and IV cannula. AV fistulas are usually formed at radial artery but can also be formed at brachial artery. AV fistula needs at least 1 to 2 months for maturation after formation. The
preferable sites for IV cannulation is right internal jugular and right subclavian vein. Femoral vein can also be used. Which is easy to cannulate, with less chance of thrombosis, and is better cosmetically but has high rate of infections. Ultrasonography guided cannulation is recommended to avoid complications.

Overall complication rate was 8% during cannulation in our study, whereas reported incidence is 20%. Reported complications in literature are: air embolism, thrombus formation, subclavian artery puncture, pneumothorax and brachial plexus injury. The complications seen in our series were haematoma in 10 patients (1.9%), cardiac arrhythmias in 5 (0.9%), pneumothorax in 5 (0.9%) and thrombus formation in 20 (4%) patients. Complications were fortunately less in our series. In case of AV fistula only one (0.3%) patient suffered arrhythmias and 3 (0.88%) had thrombus formation.

During dialysis AV fistulae worked better. In our study haemodialysis was done for four hours although in literature 5 hours dialysis is also recommended. The haemodialysis were very smooth in cases of AV fistulae. The flow rate was easily kept above 300 ml/minute but in cases of IV cannula, it was on an average 250 ml/minute. Maximum reported blood flow rates, which can be maintained, are 487 ml/minute for cases of AV fistulae and 396 ml/minute for IV cannulas. Change of position resulted in higher venous pressure in 85 haemodialysis by IV cannulas (3%) as compared to only 20 haemodialysis by AV fistulae. The cannulas were washed to get proper blood flow. Seventy five patients (2.7%) during dialysis by IV cannula suffered shivering and fever as compared to only 10 (0.5%) in cases of AV fistulae.

Post dialysis complications of AV fistulae are comparatively fewer. These were: aneurysmal dilatation of AV fistula in 10 (3%) and, rupture of aneurysm in one (0.29%) resulting in bleeding. There was no infection at the sites of AV fistulae in our series, whereas reported incidence of infection in AV fistulae is 1.2%20. Patients of PTFE graft have higher chance of infection. In cases of IV cannulas fifty patients (9.9%) started oozing blood from the cannula site. In 10 patients the cannula had to be removed. In others pressure dressing worked well to stop the oozing. Infection at the site of cannula occurred in 50 (9.9%) patients. Staphylococcus aureus was the most common pathogen at site of infection. Femoral access is associated with higher incidence of infection. Reported evidence of infection is up to 33%. It can be avoided by daily dressing with aseptic technique. The use of plastic cannula can also lead to haemolysis but no case was seen in our series.

AV fistula, once formed, worked for years with failure rate of 2.3% only in our series. Serum urea and creatinine levels decrease more effectively in cases of AV fistulae as compared with IC cannulas, in our study. It was due to better blood flow and smooth haemodialysis with less interruption in case of AV fistulae. Haemodialysis by IV cannula is costly, as cannula has to be changed after every 4-5 dialysis. Mortality, in case of IV cannula is 0.4% but none with AV fistula, Two patients expired due to pressure of haemotoma on respiratory passages and pneumothorax.

Haemodialysis by AV fistula is a far better choice for chronic renal failure as dialysis is smooth and cost-effective with less complications, specially infection. Blood flow is maintained better and fall of serum urea and creatinine is more in case of AV fistulae. Haemodialysis should be performed by AV fistula access in patients with chronic renal failure.

REFERENCES
3. Levin N. Goals of access placement, use of catheters. AJKD: Vol 30s 1997 180-181


LIVER ABSCESS IN CHILDREN

BILQUIS LAKHANI, MOHSINA USMANI

ABSTRACT:
This is a retrospective study conducted in the department of Paediatric Medicine, N.I.C.H., Karachi from January to December 1999. Total number of patients included in this study were 33. Male to female ratio was 2.6:1. These patients were under the age of 12 years. These were diagnosed on the basis of clinical and ultrasound examination. Surgical aspiration and antibiotics was the line of management.

KEY WORDS: Liver Abscess, Children

INTRODUCTION
Liver Abscess is not an uncommon condition in children. Amoebic liver abscess in more common than pyogenic abscess in Pakistan.1,2,3 In 75% of cases abscess is single and in the right lobe of liver. Large abscesses and strategically located small abscesses cause jaundice in up to 5% of patients4. Approximately 95% of patients present with pain right upper abdomen and 80% have fever. Liver abscess commonly rupture into lungs or pleura but may rupture into peritoneum or pericardium.4 Aim of this study was to collect data related to the abscess in our setup.

PATIENTS AND METHODS
Medical record of all patients who were treated in the Department of Paediatric Medicine and Surgery, NICH, between January and December 1999 were reviewed. Data included age, sex, onset of symptoms, duration of symptoms, complications at presentation, diagnostic methods, ultrasound findings and procedures performed. The management protocol included detailed history, clinical examination, investigation, surgical and medical treatment.

RESULTS
Most of the patients presented with unexplained fever, pain abdomen, tenderness in right hypochondrium, jaundice and constitutional symptoms like malaise, headache, anorexia and weight loss. (Table I).

<table>
<thead>
<tr>
<th>Clinical Features</th>
<th>No. of Patients</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fever</td>
<td>33</td>
<td>100</td>
</tr>
<tr>
<td>Constitutional symptoms</td>
<td>33</td>
<td>100</td>
</tr>
<tr>
<td>Pain abdomen</td>
<td>25</td>
<td>75.75</td>
</tr>
<tr>
<td>Pain right hypochondrium</td>
<td>16</td>
<td>48.48</td>
</tr>
<tr>
<td>Diarrhoea</td>
<td>05</td>
<td>15.15</td>
</tr>
<tr>
<td>Jaundice</td>
<td>03</td>
<td>9.09</td>
</tr>
<tr>
<td>Respiratory symptoms</td>
<td>03</td>
<td>9.09</td>
</tr>
<tr>
<td>Signs of peritonitis</td>
<td>01</td>
<td>3.03</td>
</tr>
</tbody>
</table>

* Many patients had more than one clinical feature

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TABLE-I SYMPTOMS AND SIGNS

<table>
<thead>
<tr>
<th>Lab Investigation</th>
<th>No. of Cases</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haemoglobin gm/dl (7gm/dl) (Severe anaemia)</td>
<td>07</td>
<td>21.2</td>
</tr>
<tr>
<td>Hb 7-8gm/dl (Moderate anaemia)</td>
<td>16</td>
<td>48.48</td>
</tr>
<tr>
<td>Hb 9-11 gm/dl (Mild anaemia)</td>
<td>09</td>
<td>27.27</td>
</tr>
<tr>
<td>Hb 12gm/dl (Normal)</td>
<td>01</td>
<td>3.03</td>
</tr>
<tr>
<td>T L C (&gt; 12000/cmm)</td>
<td>17</td>
<td>51.51</td>
</tr>
<tr>
<td>L F T</td>
<td></td>
<td></td>
</tr>
<tr>
<td>S. bilirubin Increased</td>
<td>03</td>
<td>09.09</td>
</tr>
<tr>
<td>SGPT Increased</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alk. Phosphatase Increased</td>
<td></td>
<td></td>
</tr>
<tr>
<td>X-ray Chest raised right dome of diaphragm</td>
<td>04</td>
<td>12.12</td>
</tr>
</tbody>
</table>

 Investigations included liver function tests, complete blood picture, x-ray chest, ultrasound of upper abdomen, serological test for amoebiasis and culture sensitivity of the abscess (Table II).
Ultrasound was done in all the cases. Right lobe was affected in 25 patients. Average size of the abscess was 5.5x4.5 cm. Three patients had more than one abscess (two or three). Complications were pleural effusion on right side in 2 cases, and in one case it ruptured into the peritoneal cavity causing generalized peritonitis. Amoebic liver abscess was more common with positive serological test for Entamoeba histolytica than pyogenic liver abscess. There was one death due to amoebic liver abscess.

DISCUSSION
Liver Abscess is one of the important health problem in developing countries. Amoebic liver abscess is one of the most prevalent tropical liver diseases. It is more common than pyogenic liver abscess in our country. All the factors needed for the spread of amoebiasis by oro-fecal route (primary route) that is contaminated drinking water and food, poor disposal of garbage and human excreta are present.

The patients' age ranged from 18 months to 12 years. Most of the children were between 4-6 years; similar age group is shown by others. Male are more commonly affected but ratio of male to female is less as compared to other studies. In our study children presented with fever, abdominal pain and tender hepatomegaly, which is comparable to study of Memon A.S. and Memon J.M. Jaundice occurred in 5% of all the cases as reported by others. Anaemia and leukocytosis were common and are comparable to study of Moores'. Ultrasound is effective in identifying and locating the abscess, for aspiration and follow-up.

In conclusion, needle aspiration under ultrasound guidance may be used to treat children with large liver abscesses or those with impending rupture alongwith drug therapy to prevent complications.

REFERENCES
NEPHRECTOMY IN CHILDREN

MUHAMMAD YOUNAS AWAN, SYED WAQQAR ALI, ALTAF HUSSAIN RATHORE

ABSTRACT:
A retrospective study to see the indications and assess the outcome of nephrectomy in 21 children was carried out at D.H.Q. and Aziz Fatima Trust Hospitals, Faisalabad from January 1993 to December 1998. There were 13 (61.6%) male and 8 (37.4%) female patients and their ages ranged from 3 days to 10 years. The commonest indications for nephrectomy were Wilms’ tumour 11 (51.6%), PUJ obstruction with non-functioning kidney 4 (18.8%), multicystic kidney 3 (14.1%), pyonephrosis 3 (14.1%) and ectopic non-functioning pelvic kidney 1 (4.1%). The results in terms of morbidity and mortality are discussed. Operative complications included inferior vena cava injury in 2, intraoperative spillage of tumour in 2 and colonic injury in one case. It is observed that pre-operative chemotherapy in cases of renal malignancy and subcapsular nephrectomy in patients with renal infection avoids complications.

KEY WORDS: Nephrectomy, Technique, Children

INTRODUCTION
Common indications of nephrectomy in children are tumour, unilateral multicystic kidney, refluxing pyelonephritis, hydronephrosis, chronic pyelonephritis, hypoplastic kidney associated with ectopic ureter and non-functioning kidney due to PUJ obstruction. This study is an attempt to outline the factors responsible for nephrectomy in our paediatric population and to compare the results with the international studies. In addition it aims at outlining the difficulties encountered during diagnosis and at the time of surgery and how to minimize the operative hazards.

PATIENTS AND METHODS
A total of 21 children were evaluated for various renal disorders. The investigation profile included complete blood examination, urinalysis, blood urea and creatinine, abdominal ultrasonography, intravenous pyelography and renal scintigraphy. In certain cases prior percutaneous nephrostomy was done and descending nephrostograms were performed later. The data was collected on a proforma to record sex, age, clinical features and management.

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RESULTS
There were 13 male and 8 female patients including 2 neonates and one infant. Eight patients were below 3 years of age. Major clinical features included abdominal mass, abdominal pain, pyrexia, pyuria, hematuria, vomiting and failure to thrive. Nephrectomy was done in 10 patients with tumour, others were unilateral multicystic kidney 2, unilateral pyonephrosis 3, unilateral PUJ obstruction 2, non-functioning renal ectopic kidney one, non-improvement of function after PCN 2 and incidental unilateral renal dysplasia one case. Operative complications included inferior vena caval injuries 2, intraoperative spill over of tumour 2 and colonic perforation one. Nephrectomy alone was performed in 7, which included 3 cases of multicystic kidney, 2 cases of pyonephrosis, one case of PUJ obstruction and one case of ectopic kidney. In 2 cases contralateral pyeloplasty and in one case contralateral pyelolithotomy were performed. In one case of pyonephrosis, right sided colon was injured inadvertently and colostomy had to be done. Nephrectomy for renal malignancy was performed in ten cases; 6 were right sided and 4 were left sided tumours. Postoperative complications comprised of wound sepsis 4, wound dehiscence 2, renal failure one, pneumonia one and recurrence of tumour one. Three patients died, two as a result of malignant process and one as a result of pyonephrosis and septicemia.
DISCUSSION
Admason et al in 1992 reviewed a total of 85 children who underwent total nephrectomy. Disease pattern in his study included renal tumour (35%), pyelonephritis (40%), multicystic renal dysplasia (19%), obstructive uropathy (7%) and renal vein thrombosis (12%). Our study showed that renal tumour (47.6%) was the principal cause of nephrectomy followed by pyonephrosis (14%). A kidney was declared non-functioning if it had less than 15% of function documented by the DTPA & DMSA scan. The scan was repeated at 3 months interval in the two cases with congenital PUJ obstruction. Renal scintigraphy has a crucial role in making up the decision of nephrectomy in a patient with renal disease. Nephrectomy should be performed if differential function is less than 10% of ipsilateral kidney measured by DMSA scan.

Kallendraft and Wallin performed 22 heminephrectomies and one total nephrectomy in children with severely deteriorated renal function in duplex renal system with function between 2% and 14% on DMSA scintigraphy. Preliminary percutaneous nephrostomy (PCN) is advised in patients with hydronephrosis when there is doubt in diagnosis or in case of impaired renal differential function on DMSA scan (between 16-30%) and in bilateral cases presenting with renal failure. We performed bilateral PCN in two cases of PUJ obstruction who presented with renal failure. Helou and Schmitt concluded that preliminary PCN in severe form of PUJ obstruction confirms the diagnosis and shows functional value of the kidney. Only 45% children with severe PUJ obstruction underwent nephrectomy after performing PCN.

Multicystic renal dysplasia is the most common cystic disease in infancy and the second most common cause of neonatal abdominal mass representing extreme degree of renal hypoplasia. The management of multicystic kidney is controversial because complete involution was observed in 48% and decrease in size in 35% of cases. It is advised that nephrectomy should not be performed in typical cases, but regular follow-up of these patients is necessary because of the potential complications of hypertension, infection and malignancy. If these complications occur in children with suboptimal follow up, nephrectomy is treatment of choice. We performed nephrectomy in three children due to multicystic renal dysplasia; one due to incidental finding during exploration for congenital intestinal obstruction. Infection in non-functioning kidney and suboptimal follow up were justification for removing kidney in other patients because in our setup parents do not visit regularly for follow up of their children due to illiteracy and ignorance about the magnitude of the problem.

In our study, 3 (14%) had both kidneys involved in some pathology and in the remaining 18 (86%), only one kidney was involved in each. In the former group, better functioning kidney (revealed by DMSA scan) was operated first, in order to improve its function. The procedures included pyeloplasty in two and pyelolithotomy in one case. DMSA Scan was repeated at an interval of 1 to 3 months and on evidence of good function, the non-functioning (less than 15%) contralateral kidney was removed. In the latter group (86%), where the pathology was limited to only one kidney, the diseased kidneys were removed.

Operative approaches in these patients were transperitoneal in 66% and subcostal renal extraperitoneal in 34% cases. Open operative removal of kidneys is the gold standard method available everywhere in the world. Though newer techniques of nephrectomy are in practice in the West, laparoscopic nephrectomy results are comparable in outcome with open surgery. Both transperitoneal and retroperitoneal approaches for the laparoscopic nephrectomy for benign renal disease have been described. Laparoscopic nephrectomy in children is feasible and associated with minimal blood loss, about 10 to 60 ml in different cases, low morbidity and no intraoperative adhesions, less perioperative pain and cosmesis. These children can be discharged 48 hours after surgery and can return to school in the first postoperative week. These procedures should be performed by an experienced laparoscopic surgical team to reduce the complications. The disadvantages of this approach are longer operative time, about 3-4 hours, its availability only at specialized centres, and intra-operative costs of surgery which is greater than that with the open surgery. The role of laser for partial nephrectomy in children who have bilateral Wilms’ tumor, duplicated system with obstructive non-functioning upper pole moieties have been reported. Operative complications were more common in patients with renal malignancy and pyonephrosis. This was because of the fact that in both the situations, the kidneys were friable or adherent to adjacent structures.

Operative complications reported are 8% in children in whom nephrectomy has been done. These complications have been small bowel occlusion (3.7%), and tumour rupture (2.8%). The latter can be avoided by preoperative chemotherapy in Wilms’ tumour. These most common operative complications for nephroblastoma is small bowel occlusion, 6.9% as reported in one study.

One of our patients with unilateral nephrectomy for Wilms tumour died of renal failure postoperatively. The cause of renal failure could not be detected in this child. The most common aetiology of renal failure found in literature was bilateral nephrectomy for recurrent bilateral Wilms tumour, another was radiation nephritis. To avoid such complications, subcapsular nephrectomy is advised over...
Nephrectomy in children

the classical nephrectomy where the kidney adherent to surrounding structures because of chronic renal infection and previous surgery.  

REFERENCES


ABSTRACT:
A comparative study was conducted in the Department of Physiology, Basic Medical Sciences Institute, Jinnah Postgraduate Medical Centre, Karachi, during the period between January 1999-May 1999. A group of 100 volunteer male subjects (50 control and 50 cotton mill workers) apparently healthy with age ranging from 20-60 years were selected randomly from general population and a textile mill at Karachi. The pulmonary function, Maximal voluntary ventilation, (MVV) was performed on auto spirometer (compact vitalograph). The cotton mill workers had significantly lower lung function (MVV) than control subjects. As the duration of exposure in cotton mill workers is increased, markedly impairment of MVV was observed (P=<0.001).

KEY WORDS: Maximal voluntary ventilation, Cotton mill workers.

INTRODUCTION
Cotton dust is defined as the dust present in air during handling or processing of cotton. Cotton dust is a complex mixture of many substances, including ground up plant matter, fibres, bacteria, fungi, soil pesticides, non-cotton plant matter and other contaminants, which may have accumulated during growing, harvesting, processing and storage. Cotton is derived from Arabic word "quotn". Cotton plant belongs to genus gossypium of the family Bombacaceae in order Malvales.

People exposed to dust from cotton, flax and soft hemp may develop mill fever, impairment of respiratory function, chest tightness, byssinosis, chronic bronchitis and hyperactive airways. A well documented change gradual decrease, over the working day, in airway flow caused by bronchoconstriction. It has also been suggested that higher the concentration of cotton dust and longer a person is exposed to it, higher is the risk of developing byssinosis and chronic bronchitis, bronchial asthma.

The maximal voluntary ventilation (MVV) was formerly called maximal breathing capacity (MBC). Normal MVV is 125-170. It/min. It depends upon the muscular force, compliance of the thoracic wall and lungs and on the airway resistance. MVV is profoundly reduced in patients with emphysema, and airway obstruction. Bronchial constriction in asthma diminishes maximal voluntary ventilation (MVV).

MVV reflects function of the entire ventilatory apparatus and there is a very close correlation between MVV and FEV1. The predicted MVV can be quickly estimated by multiplying the predicted FEV1 value by 40. When FEV1 is decreased MVV is also decreased. This relation is found not only in normal subjects but also in patients with chronic airflow limitation and in patients with pulmonary restrictive disorders’ MVV is influenced greatly by non-pulmonary factors such as motivation, sensorium, muscular force and endurance.

SUBJECTS AND METHODS
Fifty, apparently healthy male control subjects, were selected from the local population of Karachi, with age ranging from 20-60 years. Matched control subjects of similar socio-economic group, who had never been exposed to cotton dust were selected. Cotton mill workers were selected randomly from different departments of a textile mill at Karachi.

Subjects with gross abnormalities of vertebral column, thoracic cage, neuromuscular diseases, known cases of malignancy, diabetes mellitus, tuberculosis, bronchial...
Maximal voluntary ventilation in cotton mill workers

Maximal voluntary ventilation in cotton mill workers

asthma, hypertension, ischaemic heart disease and subjects, who had undergone abdominal or chest surgery were excluded from the study. The MVV was performed on auto-spirometer (Compact Vitalograph) at a fixed time of the day to minimize diurnal variations. The apparatus was calibrated daily and test was performed at 17°C-37°C room temperature. After detailed history and examination, subjects were explained the whole procedure. The test was performed in standing position. Nose clip and separate mouthpieces were used for individual subjects. The subjects were asked to carry out the following maneuver:

1. Hold the breathing tube with both hands.
2. Take deep breath and fill the lungs completely.
3. Put the mouthpiece in the mouth and breath out into the instrument as rapidly and as completely as possible.
4. Repetition of the test was done 5 times after adequate test. The computer in autospirometer self selected the highest value and gave the print out immediately with measured data along with the predicted normal values.

RESULTS

Table-I shows comparison of MVV between control and cotton mill workers. The cotton mill workers had significantly lower MVV than control subjects (P<0.001). The effect of cotton dust exposure was further determined on the basis of duration of exposure (Table-II). As the duration of exposure in cotton mill increases, markedly impairment of MVV was observed. There was no significance different is observed in MVV between control and cotton mill workers during the period of <3 years employment (exposure). The MVV was decreased during the 3-5 years of employment in cotton mill workers (P<0.05), but these values are further decreased and results are highly significant in cotton mill workers as compared to the controls during 6-8 years of employment (P=0.01) and more so in more than 8 years (P<0.001) exposure.

DISCUSSION

Cotton mill workers have lung function losses greater than those of Controls, suggesting that exposure to cotton dust in mills cause deterioration in lung function. This effect of cotton dust is confirmed in many studies including our study. Cotton dust exposure is known to produce and maintain narrow airways. It provides an excellent agent, which effect airway functions. Gandevia and Milne in 1965 had demonstrated the influence of duration of exposure in cotton mill workers and found that the FEV1 was significantly decreased in employees who had between one to ten years of exposure. Merchant et al, have found that greater the exposure of workers to cotton dust greater will be the impairment of pulmonary functions. Kitchou and Anderson have measured the FVC, FEV1, FEV1/FVC and MMFR in subjects exposed to cotton dust and showed that the FVC, FEV1, FEV1/FVC and MMFR in cotton mill workers were statistically significantly decreased. Noweir et al, reported reduction in pulmonary function with increase in duration of exposure in cotton mill.

Our results confirm that the cotton mill workers continue to have decreased lung function (maximal voluntary ventilation). This impairment of lung function is markedly effected with increased duration of exposure in cotton mills.

REFERENCES

3. Rylander, R., Schilling, R.F., Pickering, C.A.C., Rooke, G.B. and Jacobs, R.R. Effects after acute and chronic exposure...


RUKHSHANA KHURSHID, NASIM AKHTER* AND MUMTAZ BEGUM

ABSTRACT:
Cytotoxic T lymphocytes and B lymphocytes play an important role in immunity control of breast cancer. Amino acid analysis difference shows the rate of the activity. Sequence homology with other immunoglobulins and of other species show its immunity response and evolutionary relationship with other species. 3D structure shows that antigen binds to that site of hypervariable region of IgG whose shape and size is determined by amino acid sequence. In contrast Fc fragment have branched oligosaccharides and this sugar can form hydrophobic interaction of aromatic amino acid of antigen and facilitate secretion from antibody producing cells. It was found in the study that charge distribution was disturbed in breast cancer patients.

KEY WORDS: Immunoglobulin G, Breast cancer, Amino acid analysis, Sequences

INTRODUCTION
Breast cancer is the leading cause of cancer mortality in women. The cellular events that result in cancer are poorly understood. The carcinogenic process occurs in one of the two ways: by clonal proliferation or by progressive alteration of cellular growth from a reversible premalignant stage to an irreversible malignant stage. Progression is important in considering the possible role of immune control of cancer cells. Cytotoxic, killer T lymphocyte and B lymphocytes play an important role in the immune control. Cytotoxic T lymphocytes, through its receptor recognizes one of the epitopes on a desired cell, bind to epitopes and destroy the cell before more viral particles can be generated, whereas B lymphocyte give rise to cells that secrete all classes of circulating immunoglobulin molecule (IgM, IgG, IgA, IgD and IgE).

The capacity of antibody molecules themselves to specifically regulate immune response by antibody feedback mechanism is well documented. Deficient B-cell function may be important in the pathogenesis of the immune deficiency. Quantitative measurement of immunoglobulin G (IgG) is useful in a variety of clinical conditions that involve the assessment of immunocompetence and function, response to immunization abnormality and neoplastic proliferation of lymphocytes.

Purpose of study: To compare the results of amino acid analysis, sequential distribution and 3D structure of Immunoglobulin G in normal and breast cancer patients.

MATERIALS AND METHODS
Isolation, purification and characterization of IgG from serum of both normal subjects and breast cancer patients were done. Techniques used for this purpose were electrophoresis and chromatography. Its biological importance is determined by identifying antibacterial, antifungal and antitumoral activity of IgG.

Amino acid analysis.
Purified IgG from serum of normal subjects and breast cancer patients was dried in vacuum and analyzed on automatic amino acid analyzer (Biotronic LC6001, Germany).

Sequence homology
Multiple sequence alignment of IgG was done with IG Gamma -1-2-3-4, chain C region as well as with Gamma chain C region of Rabbit, rat and mouse. Amino acid sequence data of purified protein obtained from BLITZ service. 3D structure was taken from Protein data bank file.
RESULTS
Amino acid composition of IgG from serum of normal compared with amino acid composition of IgG from serum of breast cancer patient (Table I and II).

<table>
<thead>
<tr>
<th>Amino acid</th>
<th>Concentration(nmol)</th>
<th>No. of amino acid</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asp</td>
<td>0.5</td>
<td>1.0</td>
</tr>
<tr>
<td>Thr</td>
<td>28.7</td>
<td>58.0</td>
</tr>
<tr>
<td>Ser</td>
<td>14.2</td>
<td>28.0</td>
</tr>
<tr>
<td>Glu</td>
<td>28.6</td>
<td>58.0</td>
</tr>
<tr>
<td>Pro</td>
<td>67.2</td>
<td>134.0</td>
</tr>
<tr>
<td>Gly</td>
<td>39.43</td>
<td>78.0</td>
</tr>
<tr>
<td>Ala</td>
<td>17.5</td>
<td>36.0</td>
</tr>
<tr>
<td>Val</td>
<td>VL</td>
<td>VL</td>
</tr>
<tr>
<td>Cys</td>
<td>36.5</td>
<td>74.0</td>
</tr>
<tr>
<td>Met</td>
<td>VL</td>
<td>VL</td>
</tr>
<tr>
<td>Ile</td>
<td>5.4</td>
<td>10.0</td>
</tr>
<tr>
<td>Leu</td>
<td>8.7</td>
<td>18.0</td>
</tr>
<tr>
<td>Tyr</td>
<td>4.2</td>
<td>8.0</td>
</tr>
<tr>
<td>Phe</td>
<td>14.1</td>
<td>28.0</td>
</tr>
<tr>
<td>His</td>
<td>14.4</td>
<td>28.0</td>
</tr>
<tr>
<td>Trp</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>Lys</td>
<td>28.6</td>
<td>58.0</td>
</tr>
<tr>
<td>Arg</td>
<td>9.35</td>
<td>18.0</td>
</tr>
</tbody>
</table>

ND: Not determined
VL: Very low

This shows variation in the concentration of different amino acid groups especially the charge amino acids so charge distribution is being disturbed in a way decreasing the ionization and increasing the hydrophobicity or non-polarity, which result in compact folding of protein.

TABLE II
AMINO ACID ANALYSIS OF N-TERMINAL BLOCKED IgG FROM SERUM OF NORMAL SUBJECTS

<table>
<thead>
<tr>
<th>Amino acid</th>
<th>Concentration(nmol)</th>
<th>No. of amino acid</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asp</td>
<td>60.00</td>
<td>86.0</td>
</tr>
<tr>
<td>Thr</td>
<td>26.5</td>
<td>39.0</td>
</tr>
<tr>
<td>Ser</td>
<td>VL</td>
<td>VL</td>
</tr>
<tr>
<td>Glu</td>
<td>157.90</td>
<td>226.0</td>
</tr>
<tr>
<td>Pro</td>
<td>15.90</td>
<td>23.0</td>
</tr>
<tr>
<td>Gly</td>
<td>45.88</td>
<td>66.0</td>
</tr>
<tr>
<td>Ala</td>
<td>03.29</td>
<td>04.0</td>
</tr>
<tr>
<td>Val</td>
<td>VL</td>
<td>VL</td>
</tr>
<tr>
<td>Cys</td>
<td>04.00</td>
<td>06.0</td>
</tr>
<tr>
<td>Met</td>
<td>VL</td>
<td>VL</td>
</tr>
<tr>
<td>Ile</td>
<td>11.30</td>
<td>16.0</td>
</tr>
<tr>
<td>Leu</td>
<td>19.70</td>
<td>28.0</td>
</tr>
<tr>
<td>Tyr</td>
<td>06.00</td>
<td>09.0</td>
</tr>
<tr>
<td>Phe</td>
<td>01.69</td>
<td>01.0</td>
</tr>
<tr>
<td>His</td>
<td>13.00</td>
<td>19.0</td>
</tr>
<tr>
<td>Lys</td>
<td>67.65</td>
<td>97.0</td>
</tr>
<tr>
<td>Arg</td>
<td>05.40</td>
<td>07.0</td>
</tr>
</tbody>
</table>

ND: Not determined
VL: Very low

This may hide the active site of IgG inside the core and the individual becomes more susceptible to disease.
involved in formation of interchain disulphide bond. Hinge disulphide bond (Cys 229) in IgG imposes a constraint that provide a rationale for the large interstitial spaces between the two CH2 domains. All IgG contain carbohydrate (N-acetyl glucoseamine, mannose, galactose and N-glycolyglycaminic acid) in amount varying 3-12% by weight. It is associated with the C-region of H chain and coatbound to aspartate, serine or threonine side chain. A group of workers 8 reported that the role of carbohydrate is to increase the solubility of IgG, conferring protection against catabolic degradation and facilitate secretion from antibody producing cells. Fig 2 shows the sequence alignment of Fc region of human IgG with IgG sequence of other species like rabbit, rat and mouse. The amino acid sequence of rabbit IgG shares the highest degree of identity (65.5%) with human IgG whereas rat and mouse show 60% and 62% homology. This show the evolutionary relationship of human IgG with other mammals.

IgG ASTKGPSVFPLAYS5KSTSSGTGTAALGCLVKDYFPEPVTWSNWS
GCRb ...KPSVFVFAPCGGETPSSTVTGLCGLKGYPEPVTWNS
GCRa AETATAPSVYAPGTALKSNMSNVLGLCGLKGYFPEPVTWNS
GCM AKTTPPSVFPLAGSQAQNTSNVMGLCGLKGYFPEPVTWNS
IgG GALTSGVHVFAPAVLGLYSLSSVTVPSSSLGTQTYICNVTNHK
GCRb GTLTNGVRGFPSVRGSGLSSSIVSTSSGSPV.TCNVASH
GCRa GALSSGHVFAPAVLGLYTLTSSVTVPSSTWP.VTVCNVASH
GCM GLSVGVSHPVFV.LYLTLSVTVPSSPRSTVTVCNVAHP
IgG PSNTKVDKVRPSKDCTHCTPPCPAPELLPSVFLFPPKPDQT
GCRb ATNTKDYKTPASTGSKPTCPFP.ELPGPSVFIFPPKPDQT
GCRa ASSTKVDKVRPRNCGGD.CPKPCICTGVSSVFIFPPKPDQV
GCM ASSTKVDKVRPDRCGCPICTPVCPS...SVIFIPPPKPDV
IgG LMVRPTFVMTVWVHDPSQKFVNYWDQGQVHAAKTPRE
GCRb LMSLTPRTVCVVDVSQDEPOFVWMVDKVPVGAETKPRV
GCRa LTTITPVTCVYVVDISQDEFEHVFSIFWVDVEHTAVQTPRE
GCM LTTITPVTCVYVVDISQDEFEHVFSIFWVDVEHTAVQTPRE
IgG QQYXSTYRVVSLSLTVHGWNLQDKYECVKSNAKLPAIPEKTI
GCRb EQYNTTFESVPILQIQHPWRLGKFGKSYNKALPAIPEKTI
GCRa EQFNSTFRSVELPHQDWLNGRTFRCYTVSAAFPSPSIPEKTI
GCM EQFNSTFRSVELPHQDWHNLGKFKCRVNSAAFPSPSIPEKTI
IgG SKAGQPQPDPQVTVVTPSREEETKNTQVSLTLCVLGKYPFSDKAV
GCRb SKTKGAPRMDPVTLPSPSRELSKSVSTVCLINFFPAHIV
GCRa SKPEQRTGQPVHYTMSKTEEMTQNVSITCMGKVFFPDDYV
GCM ...SKTKGRKPAPQYITPPKKEQMAKDKVSCLTMIDFDPEEDITV
IgG EWESIND.GEPE.NYKTPPVLSDSGSFYSFLYSKLVDSRWOG
GCRb EAWASRNPVYSEKEYNTPPIDADQSGSFYSLTKVDKSAAVDG
GCRa EWMN.GOPQENYKNTPTMTDQSFYSLKRVNKKEKWQOG
GCM ...EWMN.GOPAENYKNCPTFMNITNGSYFVYSLNVKSNWAGEG

Figure 2: Multiple sequence alignment of amino acid of human IgG with IgG of rabbit (Rb), rat (Ra) and mouse (M). Gaps are introduced to optimize the alignment.

DISCUSSION

IgG is present in the highest concentration in serum and comprises about 75-80% of gamma globulins. Most of the circulating antibodies belong to this class. IgG is composed of 4 sub classes. IgG1 (70%), IgG2 (20%), IgG3 (6%) and IgG4 (4%). These 4 classes differ from each other on the basis of antigenic difference in the heavy chain, on the number and localization of disulfide bonds as well as on the amino acid composition. IgG is a glycoprotein made up of light (L) and heavy (H) polypeptide chains. The L and H chains are subdivided into variable and constant regions. L chain consists of one variable (VL) and one constant domain (CL). Most H chains consist of one variable and three constant domains (CH). Each domain is approximately 110 amino acid long. VH and VL stippled segments represent the hyper-variable regions of sequence (H1 and L1 etc.). These, in 3-D structure, together form the antigen binding site (Fig 3). The variable region is responsible for antigen binding, whereas the constant regions are responsible for various biological functions for example complement activation and binding to cell surface receptor.

3D structure of IgG shows both Fab (CH1, VH and VL regions) and FC fragment (CH2 region). Each homology region corresponds to a compact, independently folded unit, and these are linked together by short stretches of extended polypeptide units. The pattern of folding within each domain is similar and consists of 2 b pleated sheets with anti-parallel strands. It was found that the internal disulfide bond links the 3-stranded to the 4-stranded sheet (Fig 4). The CH2 domains differ from others because branched oligosaccharides are covalently linked to their surface i.e. oligosaccharides interact mainly with hydrophobic residue of b strands (Fig 5). A group of workers 9 reported that the most compact domain-domain association occurs between CH1:CL and between CH3:CH2. It was observed10 that the loop region L1-3 and H1-3
corresponds to the hyper-variable region of amino acid sequence. It is here that antigen binds to a site whose shape and size is determined by the sequence and folding of hyper-variable region. In contrast, the pairing of CH2 domain is markedly different i.e. one arm of each of branched oligosaccharides serve to bridge the space between CH2 domain.

It was observed by crystallographic studies on the FC fragment of IgG that sugar can form hydrophobic interaction with aromatic amino acids of antigen. It was concluded by a group that the antibody specificity is essentially a shape problem involving the 3D complimen-

CONCLUSIONS
Amino acid analysis shows that the charge distribution in IgG was disturbed in cancer patients. This may hide the active site of IgG inside the core and patients become more susceptible to the disease. Sequence homology shows evolutionary relationship of human IgG with other mammals. 3D structure shows that antigen binds to that site of hypervariable region of IgG whose shape and size is determined by amino acid sequence. In contrast Fc fragment have branched oligosaccharides and this sugar can form hydrophobic interaction of aromatic amino acid of antigen and facilitate secretion from antibody producing cell.

REFERENCES
ABDUL RAHIM SIYAL, QAMAR DIN BALOUCH, SHER MOHAMMAD SHAIKH, ABDUL WAHEED SURAHIO

ABSTRACT:
Renal cell carcinoma (RCC) is the most common neoplasm of kidney in adults. The outcome of this malignancy is generally unpredictable. However factors such as age, sex, size of tumor, laterality, histological grade and metastatic status have definite impact on its biological course. This study of 77 patients is prospective in nature. Male to female ratio in the study was 1.94:1 Average age of patients was 52.55 years. Three most common presenting complaints were loin pain, hematuria and palpable lump in the lumbar region. Right kidney was more commonly involved than left. The most common histological type was clear cell variant of renal cell carcinoma. Many of the clinicopathological aspects of the tumour were comparable to other studies except age of the patients. Our patients were about a decade younger than those in other studies. Tumours in males were larger than in females in our study.

KEY WORDS: Renal Cell Carcinoma

INTRODUCTION
Renal Cell Carcinoma (RCC) has variable mode of clinical presentation with hematuria being the single most common presenting feature. Some of the patients present with the classical triad of hematuria, flank pain and palpable mass. The causative factors in the development of RCC are largely unknown, though occupational groups such as fire fighters and painters may be at increased risk of developing RCC because of their frequent exposure to known carcinogens, namely acrolein, formaldehyde, benzene and dichlorofluoromethane. The outcome of RCC is generally unpredictable. However factors such as age, sex, size of the tumour, laterality, histological grade and metastatic status have definite impact on the prognosis. The purpose of the study was to evaluate the clinicopathological status of RCC patients whose nephrectomy specimens were received in the Department of Pathology, CMC Larkana.

PATIENTS AND METHODS
A seven year prospective study of RCC was conducted in the Department of Pathology with collaboration of the Department of Surgery between January 1992 and December 1998. All tissues of renal cancer were fixed in 10% formalin and were processed routinely for preparation of haematoxylin and eosin (H&E) stained sections. The diagnosis was made on light microscopic examination according to criteria laid down by WHO5. All cases of RCC were analysed according to age, sex, size of tumours, laterality, clinical presentation and histological diagnosis. Clinical data of the patients was collected from biopsy request forms and from the Department of Surgery.

RESULTS
Break up of all types of renal cancer is shown in Table-I. Seventy seven cases of renal cancer were received during the study period. The ages of children ranged from one to six years, with average age of 3.4 years. All 50 cases of RCC were studied in a greater detail and were analysed according to clinicopathological parameters mentioned earlier. Ages of the patients ranged from 28 to 70 years, the average and median ages were 52.55 and 50.0 years respectively. The most commonly involved age groups were fourth and fifth decades (Table-II). Presenting complaints of the patients are shown in Table-III.

Classical triad was present in 10% cases. Right kidney was more frequently involved (58%) than the left (42%).
TABLE I

<table>
<thead>
<tr>
<th>Type</th>
<th>M:F</th>
<th>Average age years(range)</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Renal cell carcinoma (Renal parenchyma)</td>
<td>1.94:1</td>
<td>52.55(28-70)</td>
<td>50</td>
</tr>
<tr>
<td>Transitional cell carcinoma (Renal pelvis)</td>
<td>1.5:1</td>
<td>50.17 (45-60)</td>
<td>05</td>
</tr>
<tr>
<td>Squamous cell carcinoma (Renal pelvis)</td>
<td>2:1</td>
<td>53.33 (45-60)</td>
<td>03</td>
</tr>
<tr>
<td>Anaplastic tumours (Renal pelvis)</td>
<td>1:1</td>
<td>57.50 (55-60)</td>
<td>02</td>
</tr>
<tr>
<td>CHILDREN (17 cases)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nephroblastoma (Wilm’s tumour)</td>
<td>1.83:1</td>
<td>3.40 (1-6)</td>
<td>17</td>
</tr>
</tbody>
</table>

TABLE II

<table>
<thead>
<tr>
<th>Size (cm)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>21-30</td>
<td>33</td>
</tr>
<tr>
<td>31-40</td>
<td>17</td>
</tr>
<tr>
<td>41-50</td>
<td>17</td>
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<tr>
<td>51-60</td>
<td>17</td>
</tr>
<tr>
<td>61-70</td>
<td>17</td>
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</tbody>
</table>

TABLE III

<table>
<thead>
<tr>
<th>Complaint</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain in lumbar region</td>
<td>33</td>
</tr>
<tr>
<td>Palpable lump in lumbar region</td>
<td>17</td>
</tr>
<tr>
<td>Haematuria</td>
<td>25</td>
</tr>
<tr>
<td>Burning micturition</td>
<td>03</td>
</tr>
<tr>
<td>Pus in urine</td>
<td>02</td>
</tr>
<tr>
<td>Backache</td>
<td>02</td>
</tr>
<tr>
<td>Oliguria</td>
<td>01</td>
</tr>
</tbody>
</table>

TABLE IV

<table>
<thead>
<tr>
<th>Type</th>
<th>No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clear cell carcinoma WD*</td>
<td>25</td>
</tr>
<tr>
<td>Clear cell carcinoma MD*</td>
<td>15</td>
</tr>
<tr>
<td>Clear cell carcinoma PD*</td>
<td>06</td>
</tr>
<tr>
<td>Granular cell carcinoma</td>
<td>02</td>
</tr>
<tr>
<td>Spindle cell (sarcomatoid) carcinoma</td>
<td>02</td>
</tr>
</tbody>
</table>

sizes of the tumours ranged between 2 and 24 cm (average: 12.97 cm) in males and between 3 and 14 cm (average: 6.75 cm) in females. The overall average size of RCC was 9.82 cm. Distribution of cases according to histological type and grade are outlined in Table IV. The leading histological type of RCC was clear cell carcinoma (92%), half of which were well differentiated.

DISCUSSION

In the present study RCC was about twice as common in males as in females. Similar figures are reported from USA and New Zealand. However the two sexes are almost equally involved in Saudi Arabia. Average age of our patients (52.55 years) is more or less comparable to that of Saudi patients (50.85 years), but it is about a decade younger than that of New Zealand group (62.5 years). Most of our patients were in their fourth and fifth decades of life whereas the peak age of New Zealand group was in the sixth decade. The possible reason for this age discrepancy could be exposure of our people to some carcinogenic agents at an early age. Another possibility could be the overall low survival rate of our people due to poor socioeconomic conditions. Three most common presenting complaints of our patients were loin pain (66%), haematuria (50%) and palpable mass (34%). Similar observations were made in a Saudi series and in other literature. Tumours of the present series were some what larger (average size: 9.8 cm) than those of the Saudi group (average size: 8.02 cm). This may be due to relative delay in the diagnosis of the disease in our patients, due to poverty and lack of education. We also noticed that the tumours of men were about twice larger (average size: 12.97 cm) than those of women (average size: 6.75 cm). Further studies in this regard are required to verify this interesting feature. Brett Delahunt et al have observed that the prognosis of right sided RCC after radical nephrectomy was better than that of the left. Majority of our patients had right sided tumours (58%). Similar laterality is reported in a Saudi study. However in a New Zealand group, left side was slightly more commonly involved than the right.

Histological pattern of RCC in the present study is more or less comparable to that of a Saudi series and to other literature.

REFERENCES


TUBERCULOSIS OF THE LUMBOSACRAL JUNCTION
LONG TERM FOLLOW UP OF 12 CASES

HAMEEDULLAH BUZDAR

ABSTRACT:
Lumbosacral junction is an uncommon site for tuberculous involvement as compared to cervical and dorsal vertebrae. Twelve cases of lumbosacral tuberculosis were reviewed over a period of 3 years from January 1994 to January 1997 with follow up period of 3-5 years in the Department of Neurosurgery, Sandeman Civil Hospital, Quetta. All the patients were adult. Usual presentation was low back pain with visible kyphus deformity. Neurological involvement was rare. Four patients were treated conservatively, while 8 patients underwent surgical intervention. Both conservative and operative treatment resulted in fusion, but all patients treated conservatively ended up with kyphus and trunk shortening. They had higher incidence of back pain. Anterior debridement with bone graft resulted in low incidence of back pain and size of kyphosis. Though anterior debridement with bone fusion with a graft is a better option for treating tuberculosis of the lumbosacral junction, but it requires experience and is technically demanding. Treatment of tuberculous spondylitis at lumbosacral junction is therefore difficult and requires individual consideration.

KEY WORDS: Tuberculosis, Lumbosacral joint, Treatment

INTRODUCTION
Lumbosacral spinal junction is a relatively uncommon site for tuberculous infection and is difficult to find in the English literature. In tuberculous spondylitis fusion of the affected segment occurs after either conservative or surgical management, with a variable angle of fusion. In this study we document our experience of this disease.

PATIENTS AND METHODS
Twelve patients with lumbosacral tuberculosis were treated and followed up for 3-5 years at Sandeman Provincial Hospital, Quetta from January 1994 to January 1997. All the patients were given full course of chemotherapy for 12 to 18 months. Out of 12 patients, surgery was performed in 8 patients while 4 were treated conservatively. Those who had surgery, anterior debridement with fusion and strut graft was carried out in 6 and two patients had posterior hemilaminectomy with deridement of the disc and contiguous vertebral bodies without fusion. At review all the patients were asked about back pain, which was graded from 1 to 5 according to Moskowitz. Forward and lateral flexion movements of the spine were recorded. Neurological examination of the lower limbs and both hip joint movements were also carried out.

The lumbosacral angle was measured using the lateral view x-ray of spine between the posterior borders of vertebral bodies closest to the apex. The commonest presenting symptom was low back pain, which increased gradually with a visible kyphotic deformity of the lower back. Two patients presented with paresis of both lower limbs. Two patients had right-sided sciatica with restricted straight leg raising (SLR) and weak dorsiflexion of the right big toe. Bladder and bowel control were not affected in any case.

RESULTS
Of the 12 patients 9 were male and 3 female. Their age ranged between 25 and 40 years. Follow up period ranged between 3 to 5 years. At review, the most common presenting symptom was low back pain with a visible kyphotic deformity of the low back region (Table I). Out of 12 patients, 6 were heavy manual workers and 3 had a sedentary life. All the female patients were...
Tuberculosis of the lumbosacral junction long term follow up of 12 cases

Out of 3 female patients 2 had conceived and each gave birth to a baby.

<table>
<thead>
<tr>
<th>TABLE-I PRESENTING SYMPTOMS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Symptoms</td>
</tr>
<tr>
<td>Low back pain with kyphosis</td>
</tr>
<tr>
<td>Discharging sinus/abscess</td>
</tr>
<tr>
<td>Paraparesis of lower limbs</td>
</tr>
<tr>
<td>Right sided Sciatica</td>
</tr>
</tbody>
</table>

There was radiographic fusion across the lumbosacral junction in all the 12 patients. However 6 were fused in kyphosis from 40% to 75%. Three patients had a lumbosacral angle of 0 degree while 3 had fusion in lordosis (5 to 35°). When there was kyphosis at the lumbosacral junction, compensatory lordosis was present in the remaining mobile lumbar segments and was also seen in the lower thoracic segments. The incidence and severity of kyphosis was related to the method of treatment (Table II). One patient had lumbosacral scoliosis due to asymmetrical damage.

<table>
<thead>
<tr>
<th>TABLE-II SEVERITY OF LUMBOSACRAL KYPHOSIS RELATED TO METHOD OF TREATMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Method of treatment</td>
</tr>
<tr>
<td>----------------------</td>
</tr>
<tr>
<td>Anterior debridement with fusion</td>
</tr>
<tr>
<td>Posterior hemilaminectomy with debridement</td>
</tr>
<tr>
<td>Conservative</td>
</tr>
</tbody>
</table>

One patient had lumbosacral scoliosis due to asymmetrical damage.

Seven patients reported back pain after healing of their disease, 4 with Moskowitz grade I, 2 with grade III and one grade IV.

At the last followup, 9 patients could reach the lower shin or below on forward flexion, two could reach the knee level only. On lateral bending all the patients could reach knee level. There was no contracture of hip. The ratio of standing height to arm span was greater than or equal to 1 in 6 patients. All other patients ranged below one.

DISCUSSION

Although the incidence of tuberculosis is decreasing, spinal tuberculosis is still an important disease.14,15 The distal lumbar spine and the sacral vertebrae are uncommon sites of involvement by tuberculosis.14 Delay in diagnosis and treatment result in long term disability.6,13,18 The treatment of spinal tuberculosis is either conservative or surgical. Surgical treatment consists of debridement with or without fusion.9,10,11

Clinical presentation of patients with lumbosacral tuberculosis varies with age. Children usually present with formation of pus and subsequent sinus is a predominant feature, while in adults the usual presentation is backache and kyphus formation.9

Back pain with kyphotic deformity was the predominant feature in most of our patients, (80 %). Though neurological involvement in lumbosacral tuberculosis is rare due to wide spinal canal and presence of roots of caudal equina instead of spinal cord,2 two patients presented with paraparesis while in two radicular pain along the sciatic nerve was the predominant feature. Back pain occurred in and around the kyphotic area and later involved the whole back. There was a direct relationship between the severity of back pain, the number of vertebrae involved and the incidence of abnormal lumbosacral angle. It had no relation with the nature of job or physical activity.

Forward flexion movements were relatively restricted in all the patients with lumbosacral tuberculosis, in our series. Hip Joint movements were restricted especially in distal lumbosacral involvement. In 80 percent of patients standing height was shorter than the arm span because of shortening of the vertebral heights due to destruction.

Vertebral bodies are the most common sites of involvement in tuberculous disease of the spine.17 The extensive anterior destruction accounts for the kyphosis at the lumbosacral junction16,17 as was observed in all our patients. The severity of kyphosis can be affected by the method of treatment.3,8,12 Anterior debridement with fusion with a strut graft improved the kyphotic angle than the conservative treatment or debridement alone.5,10,11,18 Long followup revealed smaller angle of kyphosis after anterior debridement and fusion than in a more conservative approach. The kyphosis either substantially remained the same or increased in the conservative group of our patients. Posterior approach with debridement also had similar results as that of conservative group.

It has been shown that the presence of lordosis in the lumbar spine has several biomechanical advantages and is useful for functions.4,16 The normal lumbosacral angle is 10 and 35 degrees of lordosis. The increased kyphosis or hyperlordosis (less than 10 degrees) is responsible for higher incidence of pain in the lumbar spine.4 Therefore it is important to preserve the normal angle and prevent or minimize the chance of developing kyphosis during treatment.12,13,18 Conservative treatment results in surgical fusion but does not improve the degree of kyphosis or...
rather kyphosis worsens. Three of our patients treated conservatively had increase in the angle of kyphosis after fusion. Anterior debridement with fusion requires experience and was technically demanding but had better surgical outcome. It improved the degree of kyphosis or had least kyphosis after treatment. It is therefore, imperative to consider each case individually according to the severity of symptoms and radiological involvement of the vertebrae. It is also important to keep in mind the surgical experience and the availability of facilities. However every effort should be made to facilitate a good bony union and preserve normal lumbosacral angle.

REFERENCES

4. Farfan HF. The biomechanical advantages of and hip extension for upright activityin :man as compared with other anthropoids.
PREAXIAL LIMB DEFICIENCY:

A CASE REPORT

SHAHEEN MITHANI, AYESHA RASHID, KHIZER MANSOOR, M. NAEEM KHAN

ABSTRACT:
This is a case report of a patient who presented with a renal calculus and had preaxial limb deficiencies i.e. bilaterally absent radii and tibial hemimelia on both sides. Despite an obviously deformed appearance he had remarkable functional limbs. While planning treatment, it is an important consideration whether correction of deformity will maintain or compromise the existing functions.

KEY WORDS: Preaxial limb deficiency, Preservation of function, Cosmetic appearance

INTRODUCTION
Congenital abnormalities of the extremities are classified by Frantz and O'Rahilly into seven categories. Failure of formation of parts lead to transverse and longitudinal limb deficiencies. Absence of tibia and radius is a longitudinal preaxial limb deficiency. Congenital longitudinal radial deficiencies are frequently associated with other abnormalities like blood dyscrasias and cardiac anomalies; and therefore it is important to screen these patients. Congenital longitudinal deficiency of the tibia is also associated with other anomalies.

CASE REPORT
Our patient was a 1 year old male, issue of a consanguineous marriage, born at full term. He presented with a history of hematuria and recurrent attacks of urinary tract infection. On examination, he had bilaterally deformed upper and lower limbs. Investigations showed a right renal calculus. His investigations included complete blood count, renal function test, electrolytes and liver function test which were all normal.

Skeletal X-rays revealed complete absence of the radii bilaterally and bilateral tibial hemimelia, (Fig. I & II). Pyelolithotomy was done and he recovered well. Despite the obvious deformation, he could use his upper limbs well for holding his milk bottle to feed himself and was an active and playful child. (Fig. III) Because the patient was an infant, limb surgery at this stage was not considered.

Figure I This picture shows complete absence of radius, also called radial club hand. The other limb is similarly involved.
Figure I: The x-rays of legs show bilateral partial longitudinal deficiency of both tibia.

Figure II: This picture shows longitudinal limb deficiency of all four limbs. It should be noted that despite gross radial club hand deformity, the individual learns to use hands with remarkable adaptation.

DISCUSSION

Congenital abnormalities of the extremities are caused by embryological failure of development. Frantz and O'Rahilly proposed a concise and comprehensive classification based on embryological and teratologic considerations. Categories include failure of formation, failure of segmentation, duplication, hyperplasia, hypoplasia, constriction band syndrome and generalized musculoskeletal abnormalities. Longitudinal. The transverse defect involves the entire width of the limb, as in congenital amputations, whereas in the longitudinal deficiency only the preaxial (radius, tibia) or postaxial (ulna, fibula) bones are involved. The word terminal is used when all the parts distal to and in line with the deficient portion are affected whereas the term intercalary denotes the absence of a middle part with the proximal and distal portions being present. The factor responsible for congenital limb deficiency acts on the embryo between the third to seventh weeks of gestation when the limb develops and differentiates.

The absence of both radii with bilateral hemimelia of tibia has not been reported in literature to our knowledge. The incidence of radial deficiency is quoted as 1 per 100,000 live births and the incidence of tibial deficiency is quoted as 1 per million live births. Both are known to be associated with other abnormalities in musculoskeletal, cardiovascular, genitourinary, gastrointestinal and pulmonary systems. Absence of radius is usually sporadic and not genetically patterned unless associated with a syndrome like TAR or Holt-Oram syndrome. Whereas bilateral tibial hemimelia is an autosomal dominant condition.

Congenital longitudinal radial deficiency may be subdivided into three types. Type A is the least defective and type C is the complete absence of the radius. Our case had type C radial deficiency, which is the most common type. There is total lack of radial skeletal support of the carpus and the soft tissue contracture on the radial side is severe. Markedly deviated radially, the hand may form an angle of 90 degrees or more with the forearm, and when the elbow is flexed it may even lie directly against and parallel with the arm, its radial border touching the forearm. The bones of the radial ray might be missing, the humerus might be short, the capitulum is hypoplastic and ossification of the distal humerus may be delayed. It is also important to realize that radial club hand is not a simple skeletal deficiency of the preaxial side of the upper limb; there are also abnormalities of muscles and neuromuscular structures.

Based on clinical and radiographic findings, three types of congenital longitudinal deficiency of the tibia can be delineated. According to Kalamchi and Dawe, in Type 1, there is total absence of the tibia. In Type 2, which was present in our case, the distal half of tibia is absent, but the proximal part is present to a varying degree. The femorotibial articulation is well preserved, but the proximal fibula has migrated superiorly. The flexion contracture of the knee is less marked, being 25 to 30 degrees. In Type 3, the distal tibia is dysplastic, with diastasis of the distal tibiofibular syndesmosis of varying degrees.

To obtain optimal results, early recognition and early treatment are essential. If a deformity is present it should
be carefully studied and evaluated as to its type and severity, the degree of severity, the degree of disability anticipated and the course of treatment to be followed. In severe deformities, treatment may extend over a long span of time. Supervision throughout the period of growth is often necessary. The child should be evaluated as a whole, his educational potential should be determined and he should be given vocational guidance in fields that his physical disability permits.  

According to recent studies the greatest degree of independence for these patients comes not from surgical prosthetic or orthotic intervention, but from the use of simple adaptive devices and powered mobility aids if required. This was recommended after evaluating upper and lower extremity management and it was seen that upper extremity prostheses were generally rejected, as most patients were able to perform tasks by approximating themselves closely enough to an object to use their own hands. Adaptive devices for feeding, dressing and toileting were well tolerated. In the lower extremity, most affected patients eventually precluded functional ambulation, necessitating power wheel chair or motorized cart use.

In our case the child had functional hands and therefore the necessity of surgical intervention was questioned. Unfortunately, the recommended adaptive devices are not possible in our setup with very limited facilities and financial constraints and hence these patients do not become completely independent.

REFERENCES
ECTRODACTYLY, ECTODERMAL DYSPLASIA CLEFTING (EEC) SYNDROME A CASE REPORT

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ABSTRACT: We are presenting a case of Ectrodactyly Ectodermal Dysplasia (EEC) syndrome that had bilateral complete cleft lip and cleft palate, ectrodactyly, conjunctivitis and classical skin manifestations of the disease. The baby initially presented with cleft lip and palate. Examination revealed that he also had most of the features of EEC syndrome.

KEY WORDS: Ectrodactyly, Ectodermal dysplasia, Clefting syndrome

INTRODUCTION
EEC syndrome is an autosomal dominant disorder characterized by multiple abnormalities of ectodermal tissue, limb anomalies, cleft lip and palate. The babies have fair and thin skin with mild hyperkeratosis and hypoplastic nipples. The hair are light coloured, sparse and thin. The child may have blue sclera, photophobia, conjunctivitis, dacroyocystitis and lacrimal system atresia. Limb defects are in the form of syndactyly or ectrodactyly. Teeth may be absent or may be abnormal with microdontia and caries. Occasional anomalies include deafness, renal anomalies etc. EEC syndrome is a rare autosomal dominant disorder of high penetrance. The babies have a classical appearance, however if a proper diagnosis is not made, several complications can occur. These include severe keratoconjunctivitis, recurrent infections and fatal hypothermia etc. Early recognition and management of associated anomalies and complications is of paramount importance. A multidisciplinary team approach is necessary for the management of these patients.

CASE REPORT
A 3 month old child was referred to us for repair of bilateral cleft lip and cleft palate. Examination revealed a baby with scaly dry coarse skin, fine sparse hairs, muco-purulent conjunctivitis, photophobia, ectrodactyly of both upper and lower limbs and bilateral cleft lip and complete cleft of primary and secondary palate (Fig. 1). The child was diagnosed as having EEC syndrome and further investigations were performed to rule out other anomalies. Ophthalmic evaluation excluded any lacrimal ductal system atresia and local antibiotics for conjunctivitis rapidly cleared the ocular infection. Renal ultrasonography excluded any associated anomalies. Surgery for bilateral cleft lip was performed. Child had an uneventful post-operative recovery and was discharged in satisfactory condition. The child has been scheduled for cleft palate repair and reconstruction of hands and feet.

DISCUSSION
Rudiger named Ectrodactyly Ectodermal Dysplasia Clefting (EEC) syndrome in 1970. It is one of the inherited syndromes among the ectodermal dysplasia disorders and involves primarily nails, teeth and hair. The hair are light coloured sparse and wiry. The skin is dry thin with mild hyperkeratosis. Palmo-planar keratoderma is common. Blue sclera, photophobia, blepharophimosis, corneal scarring, dacryo-cystitis, entropion, trichiasis and vasolacrimal duct stenosis are common. Limb defects are common which vary from syndactyly to ectrodactyly. Isolated cases have been reported with ectrodactyly but are very rare. The teeth have anodontia, microdontia and caries. The nails are thin, pitted and striated. Occasional hypohidrosis is common in EEC syndrome due to changes in glands that are also ectodermal in origin. Among the other associated abnormalities deafness, renal anomalies and hoarseness due to anomalies of laryngeal mucosa are common. Cleft lip and palate are essential component of this syndrome, and are usually
Ectrodactyl, ectodermal dysplasia clefting (EEC) syndrome: a case report

the presenting complaint of the patient. However families have been reported with several generations having ectrodactyly and ectodermal dysplasia without cleft lip and cleft palate. Cases have also been reported having hypothalamic-pituitary insufficiency and secondary hypogonadotropic hypogonadism. Anorectal atresia is another rare association of the syndrome. Another common problem in these children is susceptibility to various infections. Some authors believe that this is the result of a compromised immune system and thymic anomalies, others however believe that this is the result of a deficient ectodermal tissue, which makes the babies prone to infection. Antenatal diagnosis of the anomaly is possible by transvaginal ultrasonography as early as 14 weeks. Decision by the parents to terminate the pregnancy is often difficult and many families decide for the normal progression of pregnancy.

The aetiology of EEC syndrome is not known, however a balanced reciprocal translocation between 7q11, 21 and 9p12 has been reported in three generations of one family. As these disorders manifest diverse features, a multidisciplinary approach is essential in management. Surgical correction is required for cleft lip and palate, syndactyly and other structural abnormalities. Many patients have dry skin and require emollients. Keratolytics may improve palmoplantar keratoderma. If scalp hair are very sparse the patients may use a wig to avoid the psychological embarrassment. An assessment of the number of teeth, that will appear in primary and secondary dentition, can be obtained with dental x-ray in infancy and should be managed in conjunction with a Paediatric dentist. All children with eye abnormalities should be managed in conjunction with an ophthalmologist. Artificial tears are essential for dry eyes to preserve corneal damage. In neglected cases, corneal perforations can occur which need intensive management. Early detection of ophthalmic complications is therefore necessary to avoid serious complications. Reconstruction procedure will be required for stenosis of nasolacrimal ducts. The limb anomalies will need correction at appropriate age. Children are prone to develop recurrent urinary tract infections. These are often attributed to the associated immune deficiency. However studies have shown normal immunological response in these children and suggest the anatomical anomalies as the cause of recurrent urinary tract infection. Surgery in babies with EEC syndrome has to be careful as these children have poor temperature control due to hypohidrosis and can very quickly become hypothermic.
In conclusion, EEC syndrome is a complex disorder of ectodermal tissue and needs a detailed evaluation for identification of involvement of different organs and tissues. A team approach is required for its management. A close liaison is necessary between the treating specialists so that complications could be avoided and kept to minimum.

REFERENCES


HASHIMOTO'S THYROIDITIS WITH PERMANENT TOXIC STATE
A CASE REPORT

TAHIR HUSSAIN, TAHAWAR ALI, YASMIN AKHTAR

ABSTRACT:
We present a case of Hashimoto's thyroiditis presenting as a nodular goitre with toxic symptoms and raised T3 & T4 levels. Her symptoms were controlled by carbamazepine and propanolol. The toxic state persisted for two years and was finally relieved by subtotal thyroidectomy. Histological examination of resected gland showed a picture of Hashimoto's thyroiditis.

KEY WORDS: Hashimoto's thyroiditis, toxic surgical resection

INTRODUCTION
Hashimoto's disease is an autoimmune thyroiditis. It is not commonly seen in Pakistan. It usually presents as a moderately enlarged thyroid gland with euthyroid or hypothyroid status. In a smaller numbers of cases, however, it presents as a toxic goitre when it is called Hashitoxicosis. The toxic state (Hashitoxicosis) is generally transient and self limiting. We present a case in which the patient suffering from Hashimoto's thyroiditis remained toxic permanently and was only relieved by antithyroid drugs followed by surgery.

CASE REPORT
A 48 years old lady, post menopausal, presented with complaints of enlarging lump anteriorly in the neck for the two years which was associated with palpitations, inability to tolerate heat, liking for cold, weakness and weight loss.

Her symptoms were controlled by Carbamazepine 10 mg and propanolol 10 mg, 8 hourly. Patient was on these medicines for 2 years. The symptoms recurred whenever she discontinued the medicines. Laboratory investigations showed raised T3, T4 and suppressed TSH. When the drugs were withdrawn size of the gland very slowly increased and did not show regression at any point.

With the passage of time, she developed pressure symptoms, as well. When she first visited us, she was on these medicines and was euthyroid. Thyroid was moderately enlarged, nodular and moved on deglutition. The diagnosis of toxic nodular goitre was made and subtotal thyroidectomy planned. Hashitoxicosis was not suspected as the disease in very uncommon. Consistency of the gland was normal and not firm as expected in Hashimoto's disease. Sub total thyroidectomy was performed. Histological examination revealed a picture of Hashimoto's thyroiditis (prominent Hurthle changes, lymphoid follicles with germinal centres and scalloping of colloid). Patient showed a smooth recovery. Following surgery, she became hypothyroid and thyroxine was started. Six months after surgery she needs to take three-tablet of thyroxine daily. She showed antithyroglobulin and antimicrosomal antibodies in her serum. She is euthyroid on regular followup.

DISCUSSION
Hashimotos thyroiditis is a rare disease, with an incidence of 0.3-1.5 per 1000 population per year. It is 10-15 times more common in females than males and commonly occurs between 30-50 years of age. It shows familial predisposition and is genetically transmitted. The disease is characterized by replacement of thyroid tissue by lymphoid tissue consisting of lymphocytes, plasma cells, immunoblasts and macrophages. The lymphoid tissue shows germinal centres. The acini are destroyed and some of the acinar cells are seen transformed into Hurthle cells with brightly eosinophilic granular cytoplasm. The autoimmune process is cell mediated and although antithyroglobulin and anti-microsomal antibodies are found in the serum, they do not cause the disease. However they help in diagnosis and in predicting the prognosis and relapses. Rubio reported higher incidence of relapse in patients with higher
antithyroglobulin and antimicrosomal antibodies. Hashimoto's thyroiditis usually present as euthyroid, but rarely it may present as toxic state, called Hashitoxicosis. Latter Modignani et al reported a study of 67 patients suffering from chronic auto-immune thyroid disease out of which 3 showed Hashitoxicosis thus making only 4.7% of the total. Carnell and Valente reported 12.6% of Grave's disease patients developed nodules which are commonly autoimmune in nature.

Hashimoto's Thyroiditis clinically presents with typical symptoms and signs of thyrotoxicosis except the extrathyroidal signs of Grave's Disease. As in this patient there were complaints of palpitation, intolerance to heat and weight loss, the patient had tachycardia but no exophthalmos, chemosis or myxoedema. Laboratory investigation show high T3 and T4 levels in the serum but iodine uptake by the gland is generally low. ESR is high. Hashimoto's disease may be associated with other autoimmune diseases like SLE, Rheumatoid, arthritis, rheumatic fever, haemolytic anaemia, purpura, myasthenia gravis or pernicious anaemia. Takahashis described pernicious anemia at the age of 9, distal renal acidosis at 5 years and encephalopathy at 12 years of age associated with Hashitoxicosis. Hashimoto's thyroiditis is also associated with increased incidence of malignant lymphoma and papillary carcinoma. Hashitoxicosis is usually transient and does not require surgery. It is treated by antithyroid drugs or beta blockers during toxic state. Supportive hormonal therapy is continued after surgery.

REFERENCES
LATE PRESENTATION OF TYPE IV SACROCOCCYGEAL TERATOMA
A CASE REPORT

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ABSTRACT:
Sacrococcygeal teratoma is the commonest solid tumor in newborns. It can be diagnosed in utero on ultrasound. A case of type IV sacrococcygeal teratoma (entirely intra-pelvic) at the age of one year, with retention of urine, is presented. Through abdomino-posterolateral approach the mass was removed completely. At followup patient is thriving well.

KEY WORDS: Sacrococcygeal teratoma. Delayed presentation.

INTRODUCTION
The presentation of commonest variety of sacrococcygeal teratoma is so typical that hardly any other condition can be confused with it. Altman et al have described four types of this tumor. The type IV is not apparent at birth, as it is entirely intra-pelvic in position. A case of type IV lesion is described in this report.

CASE REPORT
A one-year-old female baby presented with retention of urine. The patient was febrile and abdomen was grossly distended. On palpation, a barely mobile mass was found which was arising from pelvis. At per rectal examination a huge lesion was present behind the rectum, the upper limit of which could not be reached. The patient was catheterized to decompress the bladder. Ultrasound revealed a large cystic swelling in retroperitoneum and both kidneys could be clearly identified. Alpha feto protein was within normal limits.

The patient was explored electively through laparotomy incision and retroperitoneal intra-abdominal part was mobilized. The patient was then placed in prone position and through posterior sagittal approach, the mass that was attached to lower sacrum, removed with coccyx (Figures, I and II). The major portion of the mass was cystic and contained hair and cheesy material. Post-operative recovery was uneventful. The biopsy report was that of benign teratoma.

Figure I Transperitoneal part of dissection. Cystic lesion can be appreciated

Figure II Posterior sagittal approach for removal of tumor

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DISCUSSION
A teratoma is a true tumor composed of multiple tissues of kind foreign to the part in which it arises. Sacrococcygeal region is the most common extra-gonadal site of teratoma. Altman et al have classified the tumor into four types, of which Type IV occurs in less than 10% cases. In comparison to other varieties, which are apparent to variable degree on external examination, this variety is entirely intra-pelvic and its presentation differs from others. By virtue of its presence, it causes pressure symptoms related to surrounding viscera and may present with constipation, retention of urine or as an incidental mass on palpation. In females the presentation may be delayed as urinary symptoms occur late in comparison with males.

The diagnosis is easy on clinical examination and ultrasound easily shows its details. Serum alpha feto protein is a good marker, the value of which is more in followup. A raised level in postoperative period suggests malignant change. Surgical exploration is easy as the tumor has little attachment to the adjacent tissues and most of the blood supply is derived from median sacral artery. In our patient a major portion of the tumor was intraabdominal therefore, abdominal approach was used initially to mobilize the mass, following which tumor was removed through posterior sagittal approach.

The chances of malignancy increases as the age of patient advances. Luckily the nature of tumor in our patient was benign. A long term followup is essential to detect recurrence which usually occurs in the form of malignant germ cell tumor. Alpha feto protein level usually rises with recurrence. Our patient is being followed up regularly and is doing well so far.

REFERENCES
HAZARDS OF INTRA-ABDOMINAL PACKING
A CASE REPORT

JAMSHED AKHTAR, MUHAMMAD IJAZ, ZULFIQAR MEMON, ABDUL AZIZ

ABSTRACT:
Abdominal packs are sometimes used in emergency for stopping haemorrhage from injured viscus or vessels. We are reporting a case of trauma victim in whom abdominal pack was put in to control bleeding from iliac vessels. The pack caused pressure necrosis of sigmoid colon.

KEY WORDS: Uncontrolled haemorrhage, surgical packs,

INTRODUCTION
Paediatric trauma carries a significant morbidity and at times mortality.1 Especially equipped centers are needed to deal with trauma victims. At times it becomes an obligation of the attending surgeons to deal with the injuries that requires a team approach and co-operation of other surgical disciplines. Temporary abdominal packaging can be life saving in cases of life threatening haemorrhage.2 This report describes one such case.

CASE REPORT
An eight-year old child sustained multiple injuries when he fell off a moving vehicle. Child remained conscious following accident and was rushed to a nearby District Headquarter hospital. Patient sustained multiple abrasions and friction burns over trunk and both upper limbs and few lacerated wounds over chin and foot. Patient developed abdominal distension in hospital. X-ray revealed no bony injury. The said hospital is not equipped with necessary equipment and trained personnel to deal with trauma victims, but as general condition of the patient started deteriorating, laparotomy was performed. The operation lasted for more than 2.5 hours and three units of blood were transfused. Following operation patient was immediately transferred to another DHQ hospital about 25 kilometers away for possible vascular injury. The patient was then referred to Karachi from a distance of 400 kilometers to another hospital from where patient was ultimately referred to us almost 48 hours after injury. No record of previous treatment, could be found.

On examination, patient was conscious with stable vitals. Multiple abrasions and lacerations were found all over the body. A large midline surgical incision found in abdomen. A surgical pack was found protruding out of lower part of the incision. The right lower limb was edematous with weak femoral and popliteal pulse. Capillary return of toes was more than 3 seconds. The limb was relatively cold than opposite limb and movements were intact. Repeat x-ray abdomen was unremarkable. The color Doppler showed sluggish flow through femoral artery and no flow could be detected in femoral vein. Intra-abdominal vessels could not be identified because of presence of abdominal surgical pack. As we do not have vascular surgery setup, opinion from another hospital was sought. Because of economical constraints attendants refused to go to that facility. During this period it was observed that abdomen started distending and patient developed signs of peritonitis. Patient was explored through previous incision. On opening peritoneal cavity, large amount of intestinal fluid came out. A completely disrupted small bowel anastomosis was found. The pack found adherent to bowel loops and was almost filling the pelvis (Fig. 1). The sigmoid colon was lying compressed under the pack and on removal, multiple pressure necrosed areas were found. On exploring the small bowel, it was observed that major portion of the gut was removed at previous surgery. The fresh anastomosis was performed. The length of small bowel that remained was 175 cm proximal and 5 cm distal to anastomosis. The frankly necrosed areas of...
sigmoid were removed and proximal part was taken out as end stoma and distal gut closed and left inside the abdomen. A large raw was found in right iliac fossa but no active bleeding was noted. As there was intense inflammation and edema no distinct vascular pathology could be observed. Abdomen was closed following placement of drain. Post operatively patient developed wound infection. The colostomy started functioning and patient was allowed feeding orally. The edema of right lower limb gradually decreased. The patient is due for definitive surgery.

The present case highlights the importance of establishing trauma centers in every DHQ hospital with all the facilities including equipment and personnel. Another fact that came into light is importance of referral note describing what management patient received in different hospitals, as it helps in planning further treatment. We were under the impression that only vascular injury had occurred and ignored the possibility of associated trauma. The abdominal pack was left for longer duration and ideally should have been removed within 72 hours. The packing has many detrimental effects including infection, impairment of renal and mesenteric blood flow, venous return and cardiac output.

The packing should be gentle and adjacent viscera should be protected from undue pressure. The length of the pack should be tailored according to the need. The extra-long pack with undue pressure on non-injured viscera resulted in further damage in our patient. One aspect that needs appreciation is survival of the patient that resulted from the efforts of primary attending surgeons in a facility that lacks the basic needs.

REFERENCES