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# JSP

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## EDITOR'S NOTE



## MEMORIAM



## ARTICLES



## CASE REPORTS

EDITORIAL :

# MEDICAL EDUCATION: TIME FOR A CHANGE?

Over the last several years the standard of education, specially medical education has not been what it should be. There are various factors responsible for this state of affair. It is very convenient to list reasons and to blame someone or everyone for it. Finding long lasting solutions requires hard thinking and tough work, implementing these solutions is even tougher. Effective and bold decisions must be taken now to correct and turn the situation around before it is too late. There are several ways to go about it, starting from planners to students, teachers and parents. Making effecient use of modern educational tools, information technologies and teaching strategies.

Criteria like "merit", "tests" and "evaluation" are all subjective, unrealistic and easy to manipulate to the advantage of the assessor. Students are often held responsible for everything that goes wrong, thus they suffer the consequences and may sometimes retaliate in an uncivilized manner.

To rectify the situation, measures taken may not be necessarily drastic but workable and fair, transparent and acceptable to all. To achieve this the first and foremost step or rather "leap" requires change in attitudes and to design our educational system according to the modern concepts, methods and technologies, where students are taught and trained with clear objectives to make them useful and worthy citizens. Planners and educationists must at no point forget that learning and training is much more important than evaluation.

Though the entire educational setup of the country needs serious changes, Medical Education is perhaps one of the most important concern for obvious reasons. The recent trend in our society is the "rush" for establishing Medical Educational Institutions, which has been prompted by increasing demand and requirements caused by our "population explosion". Following changes are suggested in the undergraduate medical courses to improve the medical and paramedical training in our country:-.

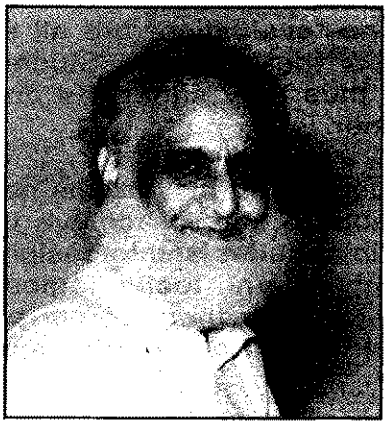
- The Basic Medical Sciences, which are Anatomy, Physiology and Biochemistry should be taught as a B.Sc. course for two years.
- Entry to MBBS should NOT be F.Sc. but B.Sc. in Basic Medical Sciences.
- Students who desire or do not get admission in the medical colleges may take up training in other disciplines like physical medicine, occupational therapy, rehabilitation, pharmacy and nursing, thus raising the standard of these very important paramedical disciplines as well.
- MBBS training should be essentially clinical for four years (including House job) with Pathology, Pharmacology, Therapeutics and other subjects evenly spread over these years.

B.Sc. Basic Medical Sciences can be taught at any institutions, thus reducing the load on hospital based institutions. To name a few advantages of this format of medical training, students for medical colleges will be better screened, they will also have their aptitude better assessed and they can change their career or even quit medical education and will still be graduates. No one will be able to open a medical college without an established full-fledged hospital and infrastructure.

**ASADULLAH KHAN**

# MEMORIAM
# DR. INAMUL HAQ SHAMI

Dr. Inamul Haq Shami, our beloved friend, companion and colleague passed away on 5th November, 1999, in Islamabad. Dr. Shami needs no introduction because of his high reputation in the profession and his efforts for the cause of medical education in the country to enhance the calibre and expertise of medical professionals.



Dr. Inamul Haq Shami was born on 10th November, 1936, in a small town near Bahawalnagar. He was eldest of a family comprising 9 brothers and 4 sisters. He did his F.Sc. from Gordan College, Rawalpindi and became the first person to have acquired F.Sc. qualification by being awarded the special scholarship of the State of Bahawalpur for the practice of Medicine as a career. Dr. Shami graduated in 1960 from Dow Medical College, Karachi, did his House Job in Orthopaedic Surgery in Civil Hospital Karachi, received postgraduate training from JPMC, Karachi and became a Fellow of CPSP after qualifying FCPS-II examination in 1972. He also obtained training at University of Illinois at Chicago on WHO Fellowship.

Dr. Shami had a very illustrious professional career starting from House Surgeon at Civil Hospital Karachi and finally rising to the post of Consultant Surgeon and Head of the Department of Surgery, Federal Government Services Hospital, Islamabad.

He was a competent dedicated, devoted and disciplined surgeon and possessed great qualities for which he was always held in high esteem not only by the doctors and students but all his colleagues, friends and patients. Dr. shami insisted on complete readiness of both doctors and para-medical staff with equipment at all times. He remained cool in emergencies, a policy that yielded success and ensured him a reputation of being an efficient and competent surgeon and good organizer. He served Surgical Department of Federal Government Hospital, Islamabad with such zeal, enthusiasm and discipline that the hospital excelled in the faculty during his tenure.

Behind his stern face, Dr. Shami, was a very kind, compassionate and cooperative person, a wise counsel for juniors, affectionate to his friends and a thorough gentleman. Laxity was the biggest sin in his lexicon and he never hesitated to admonish anyone who was a bit out of step with him. He won the heart of thousands of patients and their families for his commitment, dedication and efficiency. He was one of those who never gave hope and fought for lives of others. The secret of his success was that he kept pace with the latest research and technology and kept himself abreast of latest developments in medical sciences. He was a man of vision and worked relentlessly for the betterment of postgraduate medical education in general and for CPSP in particular.

He remained closely associated with CPSP in various capacities till the very last moment of his life. Besides being a senior Fellow, he got elected as a Councillor for the sitting Council, the position already held by him for 8 years from 1984 to 1992, for two consecutive terms. He served the CPSP as the first Regional Director and Controller of Examinations at Islamabad Centre, Honorary Faculty Member, WHO Collaborating Centre, Assessor of Papers in General Surgery, Examiner in various subjects of FCPS / MCPS. Ex-Member of faculty of Orthopaedic Surgery, Member Board of Trustees, CPSP Trust, Facilitator for different workshops conducted by the Department of Medical Education, CPSP, at Karachi, Multan, Lahore, Rawalpindi and Islamabad, Member Inspection team to inspect hospitals in the country for postgraduate training. Member of Editorial Board for Journal of Surgery, President Elect Society of Surgeons Pakistan, Ex-Vice President Pakistan Medical Association, Rawalpindi, sitting Member of Faculty of Surgery, CPSP. He has many original publications to his credit in various medical journals of Pakistan.

In him, we lost a great man who whole-heartedly served the cause of medical education meant for betterment of ailing humanity throughout his life. His departure has created a big vacuum which can hardly be filled. It is a great loss to his family and friends to the country and rather to the ailing humanity. His absence will acutely be felt by all of us and his sad demise is indeed a loss to CPSP itself.

We express our sympathy to his family in their hour of grief and pray to Almighty Allah for granting eternal peace to the departed soul and also give courage to the bereaved family to bear this terrible shock. Amen!.

**ABDUL AZIZ**

# NASAL RECONSTRUCTION WITH PEDICLE FLAPS: A REVIEW OF 77 CASES

M. SHAHID IKRAM, M. A. RAFIQUE MIRZA, ABDUL HAKIM BABAR.

*ABSTRACT:*
*A perfect nasal reconstruction still eludes the plastic surgeon. We present a study of 77 cases in which regional and distant pedicle flaps were used for reconstruction of acquired nasal defects. Patients were followed up for 6-94 months; (mean 40.5 months). Results of various flaps are compared along-with a review of literature.*

*KEY WORDS: Surgical flap, Nose deformity, Surgery plastic.*

## INTRODUCTION

Nose is the most projecting feature of the face. Being most vulnerable, it is likely to be damaged in many ways. Mutilation of nose to any degree is bound to affect the personality of the victim and consequently calls for concerted efforts to reconstruct the damaged or lost tissues. A good contour, texture and colour match are the basic requirements for nasal reconstruction[1]. The concomitant goals are restoration of an aesthetic, functional and durable nose with minimum donor site deformity.

Different reconstructive procedures including the use of skin grafts, composite grafts and local, regional and distant flaps have been described for reconstruction of nasal defects. However, only regional, distant pedicle flaps or free flaps can reconstruct major defects. Choice of reconstruction depends on the size of the defect as well as the availability of donor area. In this study we present our series of reconstruction of major nasal defects and a review of literature.

## PATIENTS AND METHODS

The study was conducted at the Department of Plastic & Reconstructive Surgery, Mayo Hospital Lahore from January 1, 1990 to December 31, 1998. Only those patients with major acquired nasal defects due to trauma, infection or excision of tumours were included in the study. 'Major defects' were defined as those which could not be treated by skin grafting, composite grafts or local transposition flaps. The different flaps used for reconstruction of major defects were conventional median, paramedian forehead temporal (Schmid's) nasolabial expanded forehead scalping forehead and arm flaps. No free flap is included in this study as required facilities are not available in our unit.

Conventional forehead flap was the preferred method of reconstruction because of its good colour match, skin texture and minimum scarring at the donor site. In the patients where this option could not be utilized due to narrow or scarred forehead, or where nasal defect was so extensive that it could not be covered with this flap, other reconstructive procedures were considered.

Expanded forehead flap was used in cases with low frontal hairline, narrow forehead or where extra tissue was needed for repair. Scalping forehead flap was utilized in patients with low frontal hairline, narrow forehead or who needed extra tissue for repair but could not afford time and expense of a tissue expander. Nasolabial flap was considered for relatively smaller defects specially in elderly individuals where the skin was lax, the defect could be covered without tension and the donor site could be closed primarily. This flap was also used in a few cases for inner lining as well. Schmid's flap was used in cases with a relatively small defect in the alar, columellar or tip regions and when the patients did not want a scar in the forehead midline area.

Correspondence:
Dr. Abdul Hakim Babar
Department of Plastic & Reconstructive Surgery
Mayo Hospital, Lahore.

Arm flap was used in some cases in which regional flaps were not available due to scarring or the defects needed extra tissue for coverage not possible with regional flaps.

All the operations were performed as elective procedures under general anaesthesia. The flaps were often modified in shape for tailor made reconstruction of the alar rim and columella. The inner lining was provided in different ways; with a folded forehead flap, with a turn down flap of skin adjacent to the borders of the defect, with a nasolabial flap from the adjacent cheek, or prefabrication with a split skin graft. Skeletal support was achieved by utilizing a conchal cartilage graft or a bone graft from the iliac crest. Donor areas were closed primarily where feasible or skin grafted where primary closure was not possible. Flaps were closely monitored during the first 24 hours for ischaemia due to kinking, venous congestion or tight sutures. Stitches were removed on the 5th or 6th day. Flap division and insetting was done after an average of 12-14 days in case of forehead flaps and three weeks in case of arm flaps.

Follow-up of patients was done monthly for the first six months, at three monthly intervals for a year and then at six monthly intervals. The aesthetic result was assessed subjectively as well as objectively and graded accordingly on a scale of 1 to 3.

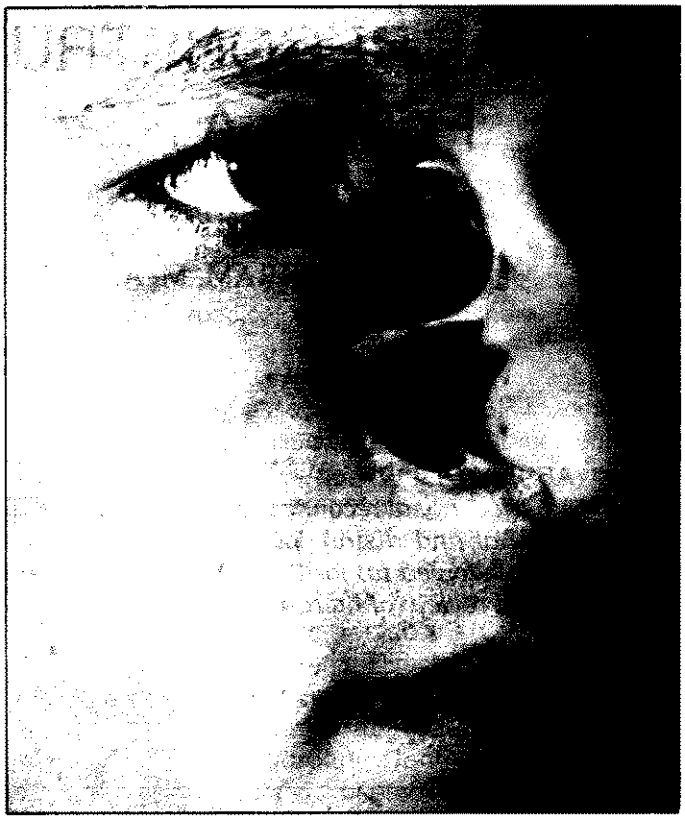


**Figure 1 (a)** A patient with post tumour excision nasal defect

**Figure 1 (b)** An inferiorly based arm flap used to reconstract the nasal defect.

## RESULTS

One hundred and forty patients were admitted for treatment of acquired nasal defects over a period of 9 years. Out of these, 77 patients (55%) had 'major' defects according to definition who needed major flap reconstruction. Male to female ratio was 1:1.4 (n= 41M and 36F). The most common cause of major defects was trauma (n=51, 66.23%). This was followed by tumours (n=15, 19.48%), post-infective defects (n=6, 7.79%) and post burn defects (n=5, 6.49%)

The results of forehead and expanded forehead flaps were good to excellent in most of the cases (Table I).

**TABLE-I AESTHETIC EVALUATION**

| Flap used | Result | | |
|---|---|---|---|
| | Excellent | Good | Poor |
| Median / Paramedian Forehead Flap | 11 | 23 | 2 |
| "Expanded" Forehead Flap | 2 | 3 | - |
| Scalping Forehead Flap | 1 | 5 | - |
| Nasolabial Flap\ | 4 | 10 | 1 |
| Schmid's Flap | 3 | 6 | - |
| Arm Flap | 1 | 4 | 1 |

The results of Schmid's flap were good to excellent in 7 cases. In 2 cases flap debunking had to be done to improve the nasal contour. The biggest drawback of scalping forehead flap was a conspicuous donor site scar; it did improve over a period of time but still caused appreciable concern to two patients. Out of 15 cases of nasolabial flaps only one became infected in a diabetic

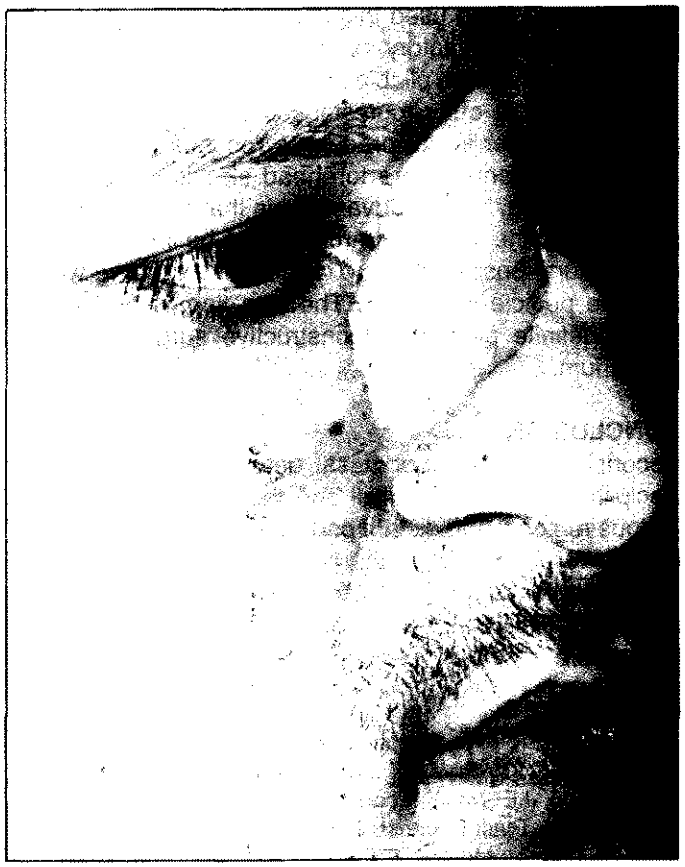


**Figure 1 (c)** Final result after division and insetting of arm falp

**Figure 2 (a)** A patient with defect of nasal tip and columella alongwith dorasal nasal scarring

patient but appropriate antibiotics and irrigation with normal saline salvaged it. Figure 1 (a) shows a patient with post tumour excision, full thickness nasal defect. An inferiority based arm flap used to reconstruct the defect. 1 (b).

**Figure 2 (b)** Frontal view after reconstruction with expanded forehead flap

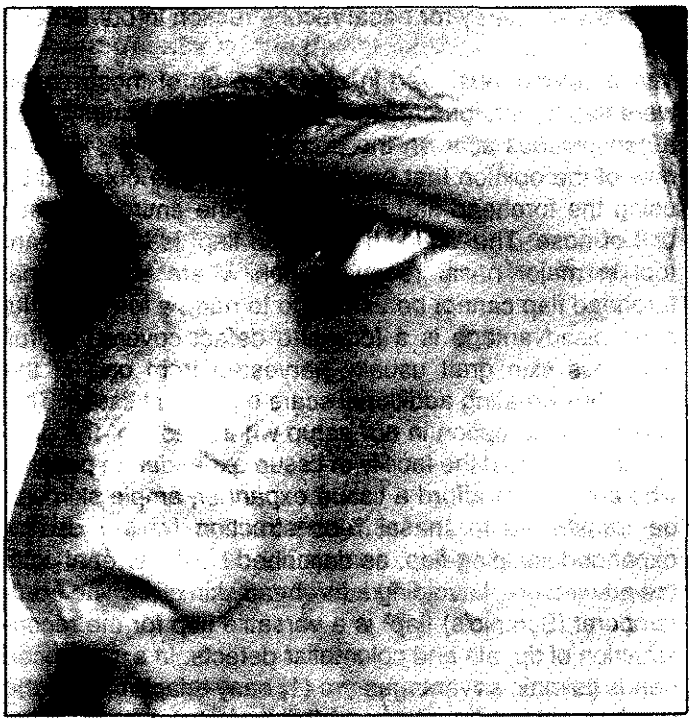


**Figure 2 (c)** Lateral view of the above

The inner lining has been provided by median forehead flap. The final result is shown in Figure 1 (c). Figure 2 (a) shows a patient with defect of nasal tip and columella alongwith dorsal nasal scarring. Figures 2 (b & c) are the post operative view of the same patient after reconstruction with expanded paramedian forehead flap.

## DISCUSSION

Reconstruction of major nasal defects still remains a challenge to the reconstructive surgeon. Historically major flap reconstruction of nasal defects are some of the earliest surgical operations described. A variety of techniques for total or sub total reconstructive rhinoplasty are available. A median forehead flap[2,3,4] provides highly vascular skin

closely identical in colour, texture and pliability to normal nasal skin and is the ideal donor site for nasal reconstruction. The resulting donor scar, especially in the older individual, is relatively inconspicuous. The flap may be elevated safely either on a single supratrochlear vessel or on an extension of the angular artery at the root of the nose[5]. The main disadvantage of the flap is the lack of adequate length to resurface the distal part of nose, especially when the frontal hairline is low-lying. An obliquely designed flap provides additional tissue to reach the base of the columella[6]. Forehead scars not in the precise midline are, however, more conspicuous. More recently, tissue expansion has been employed to provide ample tissue for any demands for nasal reconstruction yielding a wide, well vascularized flap and primary donor site closure[1]. The conventional median / paramedian flap has been the mainstay for nasal reconstruction in our series.

The problems observed in our follow-up of these cases were flap hyperpigmentation, persistent flap oedema and a conspicuous scar on the dorsum of the nose. We are now of the opinion that better results can be obtained by using the forehead flap to resurface the entire aesthetic unit of nose. The scalping forehead flap[7] is an important tool in major nasal reconstruction where conventional forehead flap cannot be used due to narrow forehead. Its main disadvantage is a forehead defect covered by full thickness skin graft usually harvested from one of the arms thus creating additional scars on normal tissue. This flap is a good option in our setup where most of the people cannot afford the facility of tissue expander. In patients who are able to afford a tissue expander, ample skin can be transferred for nasal reconstruction by the use of expanded scalping flap, as described by Zuker et al[8] with the advantage of avoiding a forehead skin graft. The fronttemporal (Schmid's) flap[9] is a versatile flap for the reconstruction of tip, ala and columellar defects. In experienced hands its main advantages are (1) easy adaptability to the requirements of the shape of the recipient site, (2) good colour match, and (3) inconspicuous scar at the donor site. However, this is a three staged procedure, flap is of limited size, and eyebrow distortion may result. We have found this to be a useful reconstructive procedure in relatively elderly patients with lax forehead skin. As this procedure is usually done in local anaesthesia, it has a definite role in patients who are unfit for general anaesthesia. Nasolabial flap[10] provides good colour match, is usually a single staged procedure and donor defect can easily be closed primarily. However only a limited quantity of skin is available. We have used this flap for inner lining of nose as well with satisfactory results. The 'Tagliacozzi' arm flap[11,12] furnishes soft, pliable medial arm skin and is a very useful option in our circumstances where the facility of free flap transfer is not present to reconstruct extensive defects in patients where forehead skin is not available due to scarring. The disadvantage is that skin is lighter in pigment and the upper extremity has to be immobilized in an unnatural position difficult to maintain for three weeks until the pedicle is divided. The arm flap, nevertheless, has a definite place in reconstructive surgeon's armamentarium.

## CONCLUSION

Reconstruction of major nasal defects is a challenging discipline, which needs skill and thorough knowledge of various regional and distant pedicle flaps. Median or paramedian forehead flap is the most suitable flap for nasal reconstruction. In patients where this flap cannot be used due to its limitations, other options can be utilized with acceptable aesthetic results.

## REFERENCES


1. Blair, V.P. Total and subtotal restoration of the nose. JAMA 1925; 85:1931-5.
2. Adamson, J.E. Nasal reconstruction with the expanded forehead flap. Plast Reconstr Surg 1988; 81:12-20.
3. Burget, G.C., Menick, F.J. Nasal reconstruction: seeking a fourth dimension. Plast Reconstr Surg 1986; 78:147-57.
4. Thomas, J.R., Griner, N. Cook, T.A. The precise midline forehead flap as a musculocutaneous flap. Arch Otolaryngol Head Neck Surg 1988; 114:79-84.
5. McCarthy, J.G. Lorenc, Z.P., Cutting, C. Rachesky, M. The median forehead flap Revisited: The blood supply. Plast Reconstr Surg 1985; 76: 866-9.
6. Baker, S.R. Regional flaps in nasal reconstruction. Otolaryngol Clin North Am 1990; 23(5):925-46.
7. Converse, J.M. Clinical applications of the scalping flap in reconstruction of the nose. Plast Reconstr Surg 1969; 43:247-59.
8. Zuker, R.M., Capek, L., de Haas, W. The expanded forehead scalping flap: a new method of total nasal reconstruction. Plast Reconstr Surg 1996; 98:155-9.
9. Meyer, R., Kesserling, U.K. Reconstructive surgery of the nose. Clin Plast Surg 1981; 8:435-69.
10. Walkinshaw, M.D., Cafee, H.H. The nasolabial flap: a problem and its correction. Plast Reconstr Surg 1982; 69:30-4.
11. Miller, T.A. Nasal reconstruction with the Tagliacozzi arm flap. In: Brent B, ed. The artistry of reconstructive surgery. St.Louis; CV Mosby, 1987: 3-9.
12. Dolomans, S., Guimberteau, J.C., Baudet, J. The upper arm flap. J Microsurgery 1979; 1:162

# ROLE OF TRANSABDOMINAL ULTRASOUND IN PARA-UMBILICAL HERNIA

TARIQ MAHMOOD, ATIQ UR REHMAN, TASNEEM IQBAL, SAEED QURAISHY,

MOHAMMAD ILYAS

*ABSTRACT:*
*In a hospital-based study, over a period of six months, 17 patients were diagnosed to have para umbilical hernia on abdominal Ultrasonography. Fourteen patients underwent surgery, which confirmed the diagnosis. Ultrasound was able to document the size of the defect which ranged from 1.0 cm to 2.9 cm. Contents of the sac were evaluated and it was found that 10 patients had omentum and seven patients had gut in the sac. Out of the seven patients containing gut loops, four patients were diagnosed as strangulation. Reducibility was also documented with certainty in 4 out of 17 patients.*

*The results showed that ultrasonography, being a non invasive easily available modality, can be used not only for detection of para umbilical hernias but also for marking the sites on skin and determining the contents of para umbilical hernias*

*KEY WORDS: Ultrasound, Para umbilical hernia.*

## INTRODUCTION

Para umbilical hernia of adult (Syn; supra or infra umbilical hernia) is a protusion through the linea alba just above or below the umbilicus. As it enlarges it becomes rounded or oval in shape with tendency to sag downwards. Para umbilical hernias can become very large. The neck of the sac is often remarkably narrow as compared with the size of the sac and the volume of its contents, which consist of greater omentum, often accompanied by small intestine and/or in addition a portion of transverse colon. In long standing cases the sac sometimes become loculated due to adherence of omentum to its fundus. Women are affected five times more frequently than men[1].

## PATIENTS AND METHODS

Patients of either sex were scanned presenting with mass in para umbilical region with or without pain. Anterior abdominal wall was also assessed in patients coming for abdominal sonography. All patients were prepared for routine ultrasound abdomen, except those who came in emergency or pregnant patients. Patients were examined with 3.75 MHz convex and 7.5 MHz linear probes on Echocee, Tosbee and Capacee. Whole abdomen was scanned initially and then para umbilical area was examined.

Correspondence:
Dr. Tariq Mahmood,
Incharge Department of Radiology, J.P.M.C., Karachi.

## RESULTS

Seventeen patients were diagnosed as having para umbilical hernia, fifteen were female and two were male. The range of age was from 35 to 52 Years. The minimum size of the defect documented was 1.0 cm and the maximum size was 2.9 cm. (Fig-1) Comments about the contents of the sac were also made in each case. In ten patients the sac contained omentum and in seven patients gut loops were seen. Out of these seven patients four were diagnosed to be having strangulation as gut wall thickening was noted along with loss of peristalsis and fluid collection around the loops. (Fig-2) In only seven out of seventeen patients hernia was found to be reducible.

Out of seventeen patients, fourteen underwent surgery, which confirmed the diagnosis. Surgery was not performed in three patients as in two patients the defect was small, measuring 1.0 cm, and contents were reducible and the clinical diagnosis was doubtful as they had thick

abdominal walls. One of the three patients was pregnant with 28 weeks gestation, so surgery was delayed.

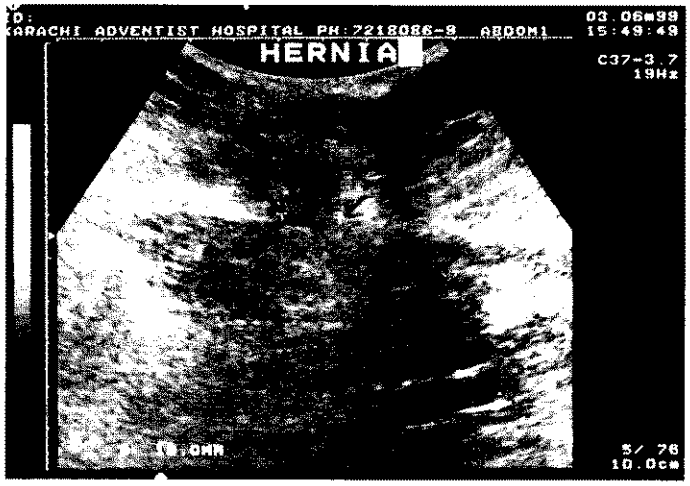



**Figure 1.** Para umbilical hernia showing 1.9 cm defect in anterior abdominal wall with herniation of mesentery.

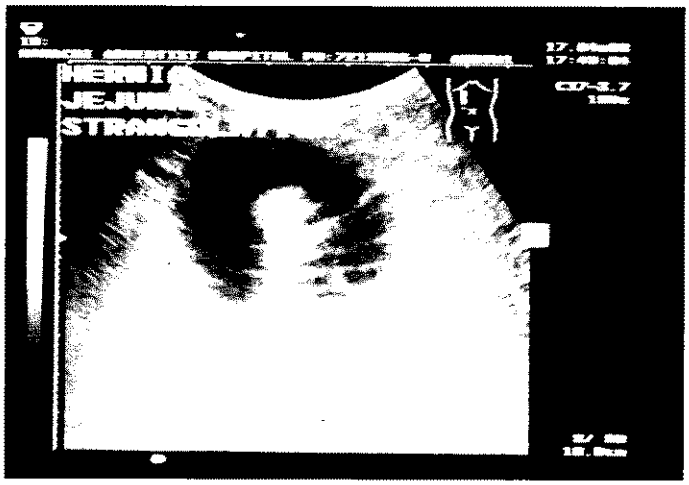


**Figure 2.** Para umbilical hernia showing strangulated jejunal loops with its valvulae conniventes.

## DISCUSSION

Anterior abdominal wall is the access for ultrasound in imaging the intra abdominal organs. One of the major roles of ultrasound is the determination of the origin of palpable abdominal masses. Masses may occur in the subcutaneous or muscle layers of the abdominal wall and may be interpreted clinically as arising from within the abdomen. If pathology originating in the anterior abdominal wall is not to be overlooked, a deliberate effort needs to be made to assess the superficial layers at the time of examination of other abdominal structures[2].

The technique required for their optimum demonstration is different from that for general abdominal ultrasound examination. It is one area of body where static scanner could retain a role because of wide field. If real time is to be used, linear array or convex transducer is ideal. High frequency transducer focused in near zone offers better resolution. The use of the split screen facility may optimize the field of view.

As we know that the superficial fascia contains a variable amount of fat. This fat is of mixed reflectivety; the fat lobules appearing relatively echo poor. It is not possible to differentiate fascias on ultrasound. The muscles of anterior abdominal wall are all-paired and consist of three layers of sheet like muscles and a fourth paired strap muscle in the midline, the rectus abdomen. Rectus abdominus reflect low level echoes occasionally with strong band like echoes from the tendinous intersections. In transverse plane the rectus abdominus appears as lens shaped and is completely ensheathed in rectus sheath which is deficient posteriorly below the umbilicus. Deep to muscle layers lie the transverse iliac fascia, it usually contains little fat and separates the muscle from the peritoneum.

Spangen advocated the use of ultrasonography in assessing ventral hernias[3]. Ultrasound is a reliable method for detecting defects of the anterior abdominal wall. The size of the defect can be easily and accurately calculated which appears as hypo echoic area in the wall, but it must be ensured that the probe is not on the umbilicus. Exact site of the defect can be marked on the skin and this is sometimes essential, as after anesthesia the fatty protusion in small hernia retracts in the abdomen and surgeons find difficulty in locating the defect[4,5]. Regarding contents, Ultrasound can accurately distinguish between omentum, mesentery and gut. Omentum or mesentery has variable appearance from hypo to hyper echoic depending upon the fat content. Small and large bowel are also differentiated on ultrasound; Reducibility can be checked and one can state with 100% certainty that the hernia is reducible, strangulated or incarcerated. Simultaneously, the state of emergency can be assessed. on B-mode scan by detecting fluid around thickened gut loops. Color Doppler will further help in assessing the blood flow in the walls of the gut. Absent flow again signifies strangulation[7]. Ultrasound in pregnant patients presenting with pain around umbilicus is very helpful where clinical diagnosis of para umbilical hernias remain doubtful. Also considering other diagnostic modalities used, like barium studies it is less time consuming, non invasive and involves no radiation hazards.

Certain pitfalls are to be kept in mind; first the umbilicus can be misinterpreted as a hernial defect. But exact localization of umbilicus on skin and position of probe above or below it can resolve the problem. Second if a strangulated bowel loop has a gas collection, this may obscure the underlying defect of hernia. This can be overcome by viewing the sac and defects from the side. Lastly, gangrenous loop gives more dirty picture, but by just prolonging examination one can be sure of diagnosis.

## CONCLUSION

Ultrasonography is a noninvasive and easily available modality not only for detection of para umbilical hernias but also for marking their sites on skin and determining contents of para umbilical hernia. Routinely, it is a clinical diagnosis by surgeons but ultrasound examination can provide valuable information, which can be of significant importance in surgery and management.

## REFERENCES


1. Bailey & Love:- Hernias Umblers . Abdominal wall by Charles V. Mum, R.C.G. Russell and Norma S. William – Bailey and Love's page 892 ELBS Edition 1995.
2. Goldberg BB. Ultrasonic evaluation of superficial masses JCW 1975: 3: 91.
3. Paul A. Dubbins, The anterior abdominal wall. IN Abdominal and general Ultrasound vol2 page 787,988 Churchill Living-Stone 1993.
4. Joseph. J. Fascia and muscles of the trunk. In: Hamilton w.j.e.d Text book of human anatomy 2nd edition 1976: 138.
5. P. Morley Abdomen, GIT and Peritoneum; In Clinical Diagnostic Ultrasound, page 301, Blackwell Scientific publications, 1985.
6. Engel J.M & Deitch E.E Sonography of the anterior abdominal wall. Am J. Roentgenol, 1981: 133, 107.
7. Fried A. M., & W.R incarcerated hernia. Ultrasound differential diagnosis. Am J. Roentgenol 1979: 133,107

# BRACHIAL PLEXUS TRAUMA : MANAGEMENT BY INTER COSTAL NERVE TRANSFER OF THE MUSCULOCUTANEOUS NERVE

HAMEEDULLAH BUZDAR

*ABSTRACT:*
*Brachial plexus injuries are common in patients with road traffic accidents and missile injuries. Management is difficult and differs according to the level of the lesion. Intercostal nerve transfer is a well-established and effective technique for irreparable brachial plexus injuries. Thirteen cases of brachial plexus injuries were treated in the Department of Neuro Surgery, Bolan Medical College Quetta by intercostal to musculocutaneous nerve transfer, without nerve graft to obtain elbow flexion; between 1994 and 1998. The results were evaluated clinically using scale developed by Wright and used by Daniels, William and Worthingham. The overall success rate with motor functions of grade 4 or more was obtained in 65 % of the patients. Factors contributing towards good results were early operation (less than 5 months after trauma), use of 3 intercostal nerves, mixed nerve to mixed nerve anastomosis, nerve repair without graft or tension and shoulder stability.*

*KEY WORDS: Nerve Trauma, Management, Nerve Graft.*

## INTRODUCTION

Intercostal nerves can be used for nerve transfer in an attempt to regain elbow flexion in root avulsion injuries or upper brachial plexus involvement. Preganglionic root injury is an irreparable brachial plexus injury[1,2]. Function is best achieved by transfer of a functioning nerve to the distal plexus[3]. Procedures for restoration of elbow flexion after brachial plexus injury include nerve grafting[1,6,7], nerve transfer[8,9] and tendon transfers[2,10,11,12]. Many surgeons perform nerve grafting for rupture of C5 and 6 roots and nerve transfer for total root avulsion. First Yeoman[13] and Seddon[4] and then Tsuyama[14] described nerve transfer of the intercostal nerve to musculocutaneous nerve without using nerve graft. Animal studies on intercostal nerve transfer have been investigated by Tomita[8] and Shoung[9].

## PATIENTS AND METHOD

Thirteen male patients, of 9 to 35 years, with brachial plexus injuries, underwent intercostal nerves transfer to the musculocutaneous nerve, over a period of 4 years from 1994 to 1998. The majority were victims of road traffic accident (7 cases), three patients had injury to the roots by missile, while 3 patients had fallen from bicycles with outstretching of the arm and concomitant injury to the nerve roots.

In all cases three intercostals nerves were used for computation. The operation was carried out under general anesthesia. A zigzag incision was made from the neck to the inner aspect of the upper arm, as described by Narakas[15]. The supraclavicular plexus was explored routinely to confirm which roots were avulsed. The phrenic nerve, if available, was cut as far distally as possible and transferred directly to the supraclavicular nerve. The spinal accessory nerve was transferred to the axillary nerve sparing the fibers to the trapezius. These two transfers were done to stabilize the shoulder joint. The semicircular incision was then extended from the wound at the anterior border of the axilla onto the infra collar fold to gain access to the intercostal nerves.

The dissection was made along the axillary vein to the subscapular vein. The mass of adipose tissue was removed from the axillary fossa which was bounded by the subclavian vein superiorly, the pectoral muscle medially, the thoracodorsal nerve posteriorly, and the subcutaneous layer of elevated skin flap laterally. This create a

Correspondence:
Department of Neuro Surgery
Bolan Medical College Sandeman Provincial Hospital, Quetta.

space for the subsequent nerve repair. The pectoral muscles were elevated without cutting or detachment. The periosteum was elevated along the lower rib margin from the nipple to the posterior axillary margin. The intercostal nerves were exposed at the anterior axillary line through the external and internal intercostal muscles. The deep central and superficial lateral branch was dissected equal. Both these branches join together proximally. Dissection was continued towards the posterior axillary line. Both the branches were cut distally and then turned to the lateral chest for suturing. Three intercostal nerves were prepared in a similar fashion for repair. Careful dissection was required for prevention of pleural tear and stretching of the intercostal nerves. The musculocutaneous nerve was found deep to the biceps muscle after dissecting the coracobrachialis muscle medially along the inner aspect of the upper arm. The nerve was dissected alongwith its origin from the lateral cord. The distal stump was mobilized distally to the biceps muscle and was transferred to the lateral chest wall for suture to the intercostal nerves. The three central branches of the intercostal nerves were sutured to the central stoma of musculocutaneous nerve while the lateral branches of the nerve were sutured to the smaller side section of the musculocutaneous nerve with 10-0 nylon. The shoulder was immobilized in adduction and the elbow in flexion by means of shoulder sling. Physical therapy and electrical stimulations were started after 3 weeks of surgery. Movements of the shoulder with lateral abduction and backward extension was avoided during physical exercises to prevent rupture of the nerve anastomosis. Clinical evaluation of the patients was performed every three months.

## RESULTS

Total root avulsion was observed in 11, while upper plexus was involved in 2 patients. The time interval between trauma and exploration varied between 3 days to 3 months. The follow up period ranged from 1 to 3 years postoperatively. Only 7 patients could be followed for longer period. (more than 2 years). Five clinical signs were observed for recovery according to the scale observed by Wright16 and used by Daniels, William & Worthingham:

- Introduction of chest pain after squeezing the biceps muscle. This is the first clinical sign observed during the postoperative course. It usually appears within the first three months.
- Proximal biceps contraction without elbow Joint Movement (M1). This sign is elicited during deep inspiration. It appears 3-6 months postoperatively.
- Distal biceps contraction. (M2). It appears subsequently after 6 months.
- Tinnel's sign; this sign is elicited during 12 months postoperatively and a tingling sensation can be elicited by purcusing the chest wall.
- Elbow flexion against gravity (M3). It appears between 12 to 18 months after surgery.
- Elbow flexion against weight (M4). This sign appears with a steady improvement every 6 months and at this stage the elbow contraction becomes independent of the respiratory movement. The radial side of forearm also becomes sensitive to light touch.

These six recovery signs are consistent and have become guidelines to predict the operative results. If any of the signs appear earlier than predicted, then the prognosis is good.

Out of 13, 7 cases were followed regularly for 4 years. Out of the remaining six, 3 could be followed upto M3 level of recovery and then were lost since they belonged to Afghanistan, while in the remaining three, two had recovered upto M2 while one disappeared after surgery. According to the clinical criteria of 7 patients followed for longer period, 4 had good results (muscle strength of grade 4 or more), 2 had fair results with a muscle strength of grade 3, while one showed improvement in the muscle power and is still being followed up[17].

## DISCUSSION

Early operation of Brachial plexus injury is recommended by most of the surgeons[4,13,14,18,19,20]. The recommended time of exposure varies from on week to 6 months[10,17,21], but this does not mean that the patients should not be operated at all beyond this deadline, especially with partial lesion or with the third or fourth degree lesions. One must consider all the facts and discuss the problem with the patient and his family[20,22,23]. If only one or two roots are avulsed, intercostals nerve transfer is not worthwhile, but if more than three roots are avulsed, then nerve transfer is indicated[3,5,6,7,19,24]. In all our cases more than three nerves roots were damaged.

The choice of using only the central branches or the complete mixed nerve transfer remains confused. Though, the transfer of only central branch, which is purely motor, is ideal due to motor transfer to the motor recipient[14], but it requires more dissection and there is possibility of damage or devascularisation with poor results[6,24]. Moreover suturing the motor branches of two intercostal nerves to the whole musculocutaneous nerve as described by Tsuyama[14] and Hara produces a discrepancy in nerve diameter. Because of this, mixed_nerve-to-mixed_nerve transfer is recommended. We therefore used three intercostal nerves with central and lateral branches, with excellent results.

The ideal number of intercostal nerves to use for nerve transfer to the musculocutaneous nerve is not decided yet. Dolence[3] used one or two, Seddon[4] used two with nerve graft (T3,T4), Tsuyama[14] and Hara and Ishii[1] used

two(T3,T4) without nerve graft. Narakas[5] used 3 to 4 intercostal nerves with nerve graft to the musculocutaneous nerve or lateral cord. Narakas[5,21] noted 1200 to 1300 myelinated fibers in one intercostals nerve while musculocutaneous nerve contained about 6000 fibers. Perhaps more intercostals nerves will provide more donor axons thereby achieving greater biceps strength[26,27].

It is not clear which intercostals nerves are best for transfer. Freilinger[26] noted no difference in motor distribution between upper and lower segments. Narakas6 reported that T4, T6 and sometimes T7 may have more axons. In this series upper intercostals nerves (T3, T4, T5) were used for nerve transfer and no difference between these nerves was found. It has also been found that T3, T4, T5 intercostal nerves can be transferred and attached with less strength, than T2, T6, T7, and are recommended for transfer.

Stabilization of the shoulder joint augments the power of the reinnervated elbow flexors[28]. We recommend reinnervation of the supraclavicular nerve and axillary nerve with phrenic and spinal accessory nerve, transfer at the same time as intercostal nerve transfer. The spinal accessory nerve was used, sparing the fibers to the trapezius muscle. This nerve transfer provides the patient with abduction of the shoulder to approximately 30°. Arthrodesis of the shoulder was not done in any case.

## REFERENCES


1. Minami M, Ishii S, Satisfactory elbow flexion in complete (preganglionic) brachial plexus injuries: produced by suture of third and fourth intercostals never to musculocutaneous never. J Hand Surg 1987; 12A:1114-8.
2. Solonen K. vastamaki M, Stron B. Surgery of the brachial plexus. Acta Orthop Scand 1984; 55:436-40
3. Dolenc VV. Intercostals meurozation of the per-ipheral nerves in avulsion of never injuries. Philadelphia: WB Saunders, 1987:425-45.
4. Seddon HJ, nerve grafting. J Bone Joint Surg 1963; 45B: 447-61.
5. Narakas AO. Thoughts on neurotization or nerve transfers in irreparable nerve lesion. Clin Plast Surg 1984; 11:153-9.
6. Narakas AO. Neurotization of nerve transfer in traumatic brachial plexus lesions. In Tubiana R, ed. The hand. Philadelphia: WB Saunders, 1988; 3:656-83.
7. Celli L, Roverta C. neuritization of brachial plexus avulsion with intercostals nevers (personal- techniques). In: Brunelli G, ed. Textbook of microsurgery. Milan: Masson, 1988:781-7.
8. Tomita Y, Tsai TM, Birns JT, Karaoguz A, Ogden L. intercostals nerve transfer in brachial plexus injuries: an experimental study. Microsurgery 1983; 4:95-104.
9. Shoung HM, Cheng HHC, Lee LS, Wu ZA, Ho DMT. Animal study on intercostals neurotization on avulsion brachial plexus injury. J Surg Assoc. ROC 1989; 22:114-9.
10. Clark JMP. Reconstruction of the biceps brachii by pectoral muscle transplantation. Br J Surg 1946; 34:180-1.
11. Tsai TM, Kalisman M, Bruns J, Kleinert HE. Restoration of elbow flexion by pectoralis major and pectoralis minor transfer. J Hand Surg 1983; 8: 186-90.
12. Akasaka Y, Hara T, Tsuyama N, Negano A. Free muscle transplantation and intercostals nerve crossing as a reconstructive procedure for elbow flexion in brachial plexus injures. Jpn J Plast Reconstr Surg 1984; 7:573-80.
13. Yeoman PM, Sedon HJ. Brachial plexus injuries: treatment of flail arm. Bone Joint Surg 1961; 43B: 493500.
14. Tsyumama N, Hara T, Nafano A. intercostals nerve crossing as a treatment of irreparably damaged whole brachial plexus. Recent developments in orthopedic surgery. Manchester: Manchester University Press. 1987:16974.
15. Narakas AO. Surgical treatment of avulsion type injuries of the brachial plexus. In Brunelli G, ed. Textbook of microsurgery Milan: Masson, 1988: 781-87.
16. Wright, N G: Muscle training in the treatment of infertile paralysis Boston Med Sci J.; 167:567 1912. Intercostal to musculocutaneous Transfer.
17. Sedel L. the results of surgical repair of brachial plexus injury. J Bone Joint Surg 1982; 64B; 54-66.
18. Miles H. brachial plexus injuries: management and results. In Terzis JK, ed. Microreconstruction of nerve injuries. Philadelphia: WB Saunders, 1987:347-60.
19. Terzis JK, Liberson WT, Maragh HA. Motorcycle brachial plexopathy. In Terzis JK, ed. Microreconstruction of nerve injuries. Philadelphia: WB Saunders, 1987:361-84.
20. Miles H, Surgical management of brachial plexus injuries. J HAND SURG 1977; 2:367-79.
21. Narakas AO. Brachial plexus injuries. In: McCa-rthy JG, ed. Plastic surgery. Philiadelphia: WB Saunders, 1990; 7:4776-816.
22. Negano A. Treatment of brachial plexus injuries Shujyustsu 1989; 43:1287-98 (in Japanese).
23. Bunnell S. restoring flexion to the paralytic elbow. J Bone Joint Surg 1951; 33A: 566-71.
24. Gu YD, Wu MM, Zheng YL, et al. microsurgic-al treatment for root avulsion of brachial plexus. Chin Med J 1987; 100:519-22.
25. Matsuda H, Nakata N, Yanai M, et al. nerve grafting for orthopaedics 1990; 21:35-42 (in Japanese).
26. Freilinger G, Holle J, Sulzgruber S. Distribution of motor and sensory fibers in the intercostals nerves. Plast Reconstr Surg 1978; 62:240-44.
27. Chang DCC, Yeh MC, Wei FC. Intercostals never transfer of the musculocutaneous nerve in avulsed brachial plexus injuries. J Hand Surg 1992; 17A; 822-8.
28. David Cheveri – Chari Chuang, et al: J Hand Surgery 1992, 17 A:822 – 8.

# MATERNAL MORTALITY: A NEGLECTED TRAGEDY

SUBHANA TAYYUB

*ABSTRACT:*
*Maternal mortality though on decline, is still alarmingly high in the developing countries. Out of 5143 deliveries over a period of 5 years from 1992 to 1996 at Dow Medical College and Civil Hospital Karachi, there were 78 maternal deaths. These maternal deaths are analysed retrospectively. The aim of this analysis was to identify the causes and contributory factors for maternal deaths. The main cause of maternal deaths was haemorrhage followed by hypertensive diseases of pregnancy. Advancing maternal age and parity, low socio-economic and poor nutritional status, lack of vigilant obstetric care coupled with delayed referral and poor facilities for transport of patients all accounted for high incidence of maternal mortality. Suggestions are also made to reduce maternal mortality.*

*KEY WORDS: Maternal mortality, Hemorrhage Hypertensive disorders of pregnancy.*

## INTRODUCTION

Woman as mother has a key position in a family, the single most important institution in nation building. Mothers are links between older and newer generations. They are the backbone of a family, looking after the newer generations in the most befitting manner. Her death during pregnancy and child birth, at a time when the children are very young, is a very severe blow to the family and the whole unit is paralyzed. Ninety five percent of children born to mothers who have died also die within one year[1].

Inspite of great advancements in medicine in the last few decades, maternal mortality is still a major worldwide challenge. It usually reflects the standard of general health of population and is associated with socio-economic deprivation. It is a sensitive indicator of health status of a country. Approximately half a million women die in the world each year as a result of complications during pregnancy and child birth and 99% of them occur in the third world. More than half of these deaths occurs in South Asia. The life time risk of maternal death for African women is one in 25 compared to one in several thousands for women in developed countries[2]. What is currently witnessed in the developing world represents the state of affairs in developed countries many years ago[3].

Maternal mortality rates in developed countries have been generally brought to less than 15/100,000[2]. Maternal mortality has fallen in New South Wales from 630 in 1935 to 11.8/100,000 births in 1991. Most developed countries have achieved such significant reduction that a maternal death is now a rare occurrence[4], while in the developing countries it still ranges from even 50 to 2000 per 100,000 life births.. Maternal mortality in Pakistan is also high as evidenced by available data, according to current estimates it is 500/100,000 live births, 25000 women dying each year of pregnancy related causes[5] It is also estimated that one in every 38 women dies from pregnancy related cause compared for example with one in every 230 women in Sri Lanka[6].

The aim of this study was to analyse various causes, to identify contributing factors responsible for maternal deaths and to assess avoidable factors. Some of the results are compared with other studies. Recommendations to reduce maternal mortality are also made. A critical review is made of all these deaths, keeping the classification made by FIGO Committee as the basis.

## PATIENTS AND METHODS

This retrospective study on maternal mortality during the 5 years period between 1st January 1992 and December

Correspondence:
Dr. Subhana Tayyub
Assoc. Prof. of Obst. & Gynae.,
Lyari General Hospital, Karachi.

1996 was carried out in Gynaecology and Obstetric Unit II Civil Hospital, Karachi during which 78 maternal deaths occurred. Civil Hospital Karachi, attached to Dow Medical College, is a government run, free hospital in the metropolitan City of Karachi, which serves as a major referral centre for secondary and tertiary care, for urban, semi-urban and rural areas with in a 50 miles radius. However, antenatal care and registration is nor selective or restricted. No patient in labour is refused admission whether booked or unbooked, referred, complicated or otherwise. Proportion of patients booked for antenatal care is approximately 20-25%.

In the presented analysis, all maternal deaths are included during the study period, whether the woman was booked or unbooked, referred in or died within minutes of arrival from home or other hospitals. The definition and classification of maternal death used in this study is that recommended by the World Health Organization, as any death of a woman during pregnancy or within 42 days after termination of pregnancy, from a cause related to or aggravated by the pregnancy or its management but not from incidental or accidental causes[7].

Records of all the patients were retrospectively analyzed in relation to age, parity, social class and booking status or referred, mode and place of delivery time interval between admission and death. Most of the patients were referred, mainly from private clinics, small nursing homes, not essentially managed by experienced medical personnels dealing with maternity cases, who referred them only when the mother developed some serious complication. Some were managed at home by a senior member of the family or by an untrained "Dai", traditional birth attendant (TBA). A large number of serious cases also come from interior of Sind specially Thatta district, and from Balochistan. These areas are inaccessible and hence the patients are brought late in shock and in moribund state. Special efforts were made to find out causes of maternal deaths amongst them. The causes of maternal mortality could be single or multi factorial.

Whenever there was more than one contributory cause, the main cause of death was recorded, clinical diagnosis was assigned for final coding but the precise pathological processes were not clear as autopsy was not performed in any case because of the social attitudes, relatives not consenting for it.

## RESULT

There were 5143 deliveries from January 1, 1992 to December 31, 1996 and there were 78 maternal deaths within that period, giving the maternal mortality rate of 15.17 per 1000 total births out of 78 patients, 55% were non booked, 45% were referred from Private Clinics and nursing homes. No deaths occurred in booked patients.

## MATERNAL AGE AND PARITY

In the present series, the ages of the patients ranged from 17-45 years (Table –I). Maximum number of deaths i.e. 66 (84.61%) were in the age group 21-40 years.

| TABLE-I | AGE WISE DISTRIBUTION OF WOMEN | |
|---|---|---|
| Age in year | No | % |
| 17-20 | 03 | 03.84 |
| 21-30 | 31 | 39.75 |
| 31-40 | 35 | 44.87 |
| >-40 | 09 | 11.54 |
| **Total** | **78** | **100.00** |

The death rate was significantly related to parity. Table II shows, parity wise distribution of maternal deaths. The largest number of patients i.e. 35 (44.87%) were grand multipara the highest parity recorded was 12.

| TABLE-II | PARITY WISE DISTRIBUTION OF MATERNAL DEATHS | |
|---|---|---|
| Parity | No | % |
| Primigravidae | 19 | 24.36 |
| 1-2 | 08 | 10.25 |
| 3-4 | 16 | 20.52 |
| 5 and above | 35 | 44.87 |
| **Total** | **78** | **100.00** |

Maternal deaths in relation to time of pregnancy is given in Table III. Most of our cases i.e. 38 (48.72%) were post partum.

| TABLE-III | MATERNAL DEATHS IN RELATION TO TIME OF PREGNANCY | |
|---|---|---|
| Relation to pregnancy | No | % |
| Antepartum | 9 | 11.54 |
| Intrapartum | 22 | 28.20 |
| Post Partum | 38 | 48.72 |
| Abortions | 9 | 11.54 |
| **Total** | **78** | **100.00** |

Nature and place of delivery are shown Tables-IV and V. Out of 78 maternal deaths, 27 (34.6%) delivered at home and 33 (42.4%) in hospital. Out of these 33 patients, 14 women (42.43%) had caesarian section 9 (27.27%)

| TABLE-IV | PLACE OF DELIVERY | |
|---|---|---|
| | No | % |
| Home delivery | 27 | 34.6 |
| Hospital delivery | 33 | 42.4 |
| Undelivered | 09 | 11.5 |
| Abortion | 09 | 11.5 |
| **Total** | **78** | **100.00** |

TABLE-V MODE OF DELIVERIES IN HOSPITAL

| Mode of deliveries | No | % |
|---|---|---|
| Spontaneous delivery | 6 | 18.18 |
| Forceps delivery | 3 | 9.09 |
| Vacuum delivery | 1 | 3.03 |
| Caesarean section | 14 | 42.43 |
| Laparotomy | 9 | 27.27 |
| **Total** | **33** | **100.00** |

patients had rupture of uterus at laparotomy, 3 patients was delivered by forceps at private maternity homes who arrived in a very serious condition and died within one hour of admission. Another patient was admitted after vacuum delivery in a private hospital with a huge infralevator haematoma and DIC in a moribund state.

## TYPE OF MATERNAL DEATHS

The deaths were classified as per FIGO classification; 92.31% were direct obstetric deaths and 7.69% being indirect obstetric deaths.

## CAUSES OF MATERNAL DEATHS

Distribution of major causes of death is shown in Table-VI. There were higher proportion of fatalities attributed to hemorrhage, hypertensive diseases of pregnancy including eclampsia, puerperal sepsis, rupture and gravid uterus and sequlae of illegally induced abortion were collectively responsible for 91% of maternal deaths. There were two deaths due to anaesthesia complication and 5 due to medical disorders. It is remarkable that there were no deaths in ectopic pregnancy.

TABLE-VI CAUSES OF DEATH

| Cause of Death | No | % |
|---|---|---|
| Haemorrhage | 26 | 33.33 |
| APH | 11 | |
| PPH | 15 | |
| Hypertesince disease of pregnancy including eclampsia | 14 | 17.95 |
| Sepsis | 11 | 14.10 |
| Rupture of uterus | 10 | 12.83 |
| Abortions | 10 | 12.83 |
| Anaesthesia complications | 2 | 2.56 |
| Medical Causes | 5 | |
| Hepatic failure | 2 | 2.56 |
| Cardiac failure | 1 | 1.28 |
| Anemic failure and transfusion reaction | 1 | 1.28 |
| Pulmonary embolism | 1 | 1.28 |
| **Total** | **33** | **100.00** |

## DISCUSSION

There is a steep decline in maternal death rates in most western countries, in Scandinavia it has fallen to 10 per 100,000 births and 7.72 in UK,[11] but it did not decline in this way in developing countries. It is at least 100 times higher than in Western Europe and North America[12]. This achievement in reduction of maternal mortality over decades in developed countries was due to interpaly of multiple factors like increased and effective antenatal. intranatal and postnatal cares. various new biochemical and biophysical methods of investigations, new diagnostic and therapeutic measures, effective management of labour[13], and better training in obstetrics and anaesthesia (London 1992).

Pakistan is one of the few countries in the world where maternal mortality is quite high (a reflection of poor maternal health services) Pakistan's population is estimated to be over 130 million, with women of child bearing age constituting about 20% of the total population. Our literacy rate is 40% and fertility rate of 5.4 is considerably higher than other large Asian countries. Only 24% of married women use contraception. High fertility is explained by the unmet need for contraception. High annual population growth rate (2.8%) impedes efforts to reduce poverty and high fertility adversely effects maternal outcomes. Health facilities are available to about 55% of population. Most of these health and MCH facilities are concentrated in urban areas. A 5 years survey[13] carried out in Pakistan showed that 70% of women received no antenatal care. Where care was received, 23% were provided by doctor, 3% by a nurse, LHV or family health worker and 4% by trained birth attendant. About 20% of births are assisted by appropriately skilled health providers. Over 5 million deliveries take place each year. Although accurate figures for maternal mortality in Pakistan are not available, an estimated maternal morbidity rate (MMR) is 340 – 500 maternal deaths for every 100,000 live births[14]. It is also estimated that one in every 38 women die from pregnancy related causes[8]. MMR is also high in other developing countries like Nigeria[15], Uganda, Bangladesh[16], India[17] and other Asian countries. The observed high maternal mortality in present analysis calls for self criticism. It is comparable to other studies conducted in various institutions of the country and it varies widely between different areas of the same region.

Haemorrhage was the predominant contributor to maternal deaths in the presented series (26/78). A further analysis of the 26 maternal deaths form haemorrhage showed 15 patients who died had post partum haemorrhage and all of them had delivered outside; 9 came with retained placenta and hemorrhagic shock, 4 patients who delivered at home, were admitted with severe PPH in severe

shock, 2 patients were referred from peripheral hospitals after forceps deliveries, one with multiple vaginal and cervical tears who died within one hour of admission, the other with a big infralevator and vulval haematoma, died due to DIC. Active management of 3rd stage of labour can prevent post patum haemorrhage.

Out of 11 deaths due to antepartum , 5 deaths were due to placenta praevia, all of them were admitted in a state of severe shock, 2 of them died undelivered. There were six deaths from placental abruption, all of them were referred from private maternity homes in a desperate condition, of these 4 developed disseminated intra vascular coagulation and other 2 deaths were due to acute rend failure.

The second most common cause of material deaths was hypertensive disorders of pregnancy, 14(17.95%) maternal deaths occurred due to this complication. Among these deaths, eclampsia was the predisposing cause in 11 patients and severe PIH in 3 cases. Out of these 11 cases of eclampsia 6 were antepartum, 3 were intrapartum, 2 were post partum eclmpsia. High incidence of PIH and eclampsia in our series is probably due to more referrals of this complication, although PIH is a disease of young primigravid deaths associated with PIH. Eclampsia in our patients occurred more commonly in the old age group and in multiparious women.

The third most common cause of maternal death was perpural sepsis. On further analysis of these 11 maternal deaths due to sepsis, 5 were due to septicemia shock, 2 died due to generalized peritonitis, one patient who died after evacuation of the uterus and colpotomy was in anaemic failure on admission. Post partum sepsis also was responsible for 2 more deaths where cause of death was acute renal failure in one case and left ventricular failure in an other case. Most of them were delivered at home by untrained TBAs who perhaps did not observe aseptic techniques.

Rupture of gravid uterus claimed 10 lives. Illegally induced abortions continue to be one of the major causes of maternal deaths, accounting for 12.83% of the total maternal deaths. The causes of death were mainly trauma and sepsis.

International data suggests that maternal mortality is decreasing in regions where the use of family planning is increasing because of consequent avoidance of unwanted pregnancies. High incidence of maternal deaths due to illegally induced abortion is related to the fact that abortions are restricted and illegal in Pakistan.

Two patients died due to anaesthesia problems accounting for 2.56% of maternal deaths during the said period while in Hyderabad[18] and Lahore[19] this figure was 2.78%, and 4.6% respectively. Pregnancy complicated by cardiac diseases, hepatic and renal problems remain our major indirect causes of maternal deaths. Pre-pregnancy counselling, good care during pregnancy, labour and puerperium are mandatory for these patients.

## CONCLUSION

In conclusion the reported maternal mortality rate is high and efforts should be made to reduce it as most of the causes are preventable.

Recommendations to reduce maternal mortality are:

- Mass education and public awareness by press, radio, television. Well-organized primary community health services, health care programmes, can improve situation by imparting adequate health education.
- Establishment of pre-pregnancy counseling clinics can reduce the incidence of maternal deaths associated with diabetes, hypertension and cardiac diseases etc.
- Expanding the coverage and quality of prenatal, delivery and post partum care.
- A fully functioning referral system ensuring quick referral and transport for life threatening emergencies.
- As the frequency of maternal deaths in a country depends not only on the risk of an average pregnancy but also on fertility rates, information provision, education and communication can create a demand for effective family planning, which will definitely lead, to a decline in maternal mortality.

## REFERENCES


1. Chen LC Gesch MC, Ahmed S, Chowdhury Ali, Mosley WH, Maternal mortality in rural Bangladesh. Studies in family planning 1974, 5,. 11: 334-341.
2. Zahr CA, Roystom E, Maternal Motility A global fact book Division of family health. Geneva:WHO, 1991.
3. Fathalla M.F. Safe motherhood. Int. J. Gynaecol Obstel 1998; 26:347-348.
4. Gutierrez H, Houdaille J. La mortalite maternellen France au. XVIII c Siecle Population 1983; 6:975-994.
5. Elliot P. The reduction of maternal and perinatal mortality in developing countries is possible. J.Coll. Phy. Surg. Pakistan 1993; 3(1) 4.
6. Maternal mortality rates FHE/852 W.H.O. 1985.
7. Jafarey S.N. Maternal mortality in Pakistan, an overview. Proceedings of a workshop on maternal and Perinatal health 1991.
8. Tinker AG, Improving women health in Pakistan. Human development network WHO and UNICEF 1998, 5-7,26.
9. World Health organization international classification of diseases, injuries and deaths, 9 the ed. Geneva 1979.
10. Unifield C.R. Vital statistics and information for obstetricians, Dewhursts Text Book of Obstetrics and Gynaecology, fifth edition page 503.
11. Duley L Maternal Mortality associated with hypertensive disorder in Africa, Asis, Latin America and Caribean Br. J. Obstetric Gynaecol 99,547-53.
12. Pandit R.D. Role of antenatal care in reducing maternal mortality Asia-Oceania. Obstet. Gynaecol 18, 1-6 1992.

13. Rukanuddin AR and Hasan KZ: Maternal and child health. In: Pakistan Demographic and Health survey 1990 / 1991. Published 1992. By National Institute of Population Studies. Islamabad, Pakistan and IRD / Macro International Inc. Columbia. Maryland USA.
14. Zaidi S, Gabb R. A manual for reproductive health training in Pakistan 1996.
15. Olalekan A, Maternal mortality a 12 years Survey at the university of Ilorin teaching hospital Ilorin Nigeria. Internal J. Gynaecol. Obst 1987;25:93-98
16. Aladdin M; Maternal mortality in rural Bangladesh; Tangail district study in. Family planning 1986; 17; 13-21.
17. Santpur. SR Maternal mortality in rural based teaching hospital Asia Oceania J. Obstet Gynaecol 1992 18, 1 31-35.
18. Farooq SM. Maternal mortality at Liaquat Medical College, Hyderabad J. Coll. Phy. Surg. Pakistan 1993; 3: 8-11.
19. Najmi R.S. Maternal mortality a hospital based study J. Coll. Phy. Surg Pak 1995 ; 5:71-4.
14. Kouris B., Cyftaki E., et al, The values of pre and post-operative scanning in the liver Echinococcosis. Br. J. Surg. 1970, 57 : 178-83.
15. Morel P, Robert J, Rohner A. Surgical treatment of hydatid disease of the liver : a survey of 68 patients, Surgery 1998 104 : 850-862.
16. Morris D. L. Surgical management of hepatid hydatic cysts. In : Morris D. L., Richards R. S. (eds) Hydatid disease. Butterworth-Heinemann, Oxford, 1992 57-75.
17. Pollick E. W. Hydatid disease of the liver Am. J. Surg. 1979, 138, 805-808.
18. Stallbaumer MF, Clarkson MJ, Richard FJ. et al. Serological diagnosis and current epidemiology of hydatid disease in England and Wales. Gut 1983, 24 A. 1996.
19. Belghiti J., Benhamous J., Houry S. et al Caustic sclerosing cholangitis. Arch. Surg. 1986 121 : 1162-1165.
20. Papadimitriou J., Mandrekas A. The surgical treatment of hydatid disease of the liver. Br. J. Surg. 1970. 57: 431-433.

# INCIDENCE OF GALL BLADDER MALIGNANCY IN PATIENTS WITH CHOLELITHIASIS

SHER MOHAMMAD SHAIKH, IQBAL AHMED SHAIKH, SIKANDER ALI MUGHAL

*ABSTRACT:*

*A prospective study was carried out in the Department of Surgery, Chandka Teaching Hospital, Larkana from January 1997 to December 1998. One hundred cases of cholelithiasis were reviewed for malignancy (80 females and 20 males). Out of these 10% patients were found to have carcinoma of the gall bladder. Their ages ranged from 41 to 69 years, with mean age of 52.2 years. Carcinoma of the gall bladder was found to be more common in the 5th decade of life. Common presenting symptoms were upper abdominal pain (100%) nausea and vomiting (80%), weight loss (50%), jaundice (40%) and anorexia (20%) Abdominal mass was palpable in two patients. Preoperative diagnosis with the help of ultrasound was possible only in three cases. Histopathologically, all the cases were adenocarcinomas. In majority of the cases, stones were mixed with cholesterol as a predominant component..*

*KEY WORDS: Gallbladder with carcinoma, biliary tract malignancy, gallbladder, neoplasm-surgery.*

## INTRODUCTION

Carcinoma gallbladder is uncommon, although not rare and is the most common biliary tract malignancy[1]. In most cases, due to lack of specific symptoms and signs, the diagnosis is late. Gallbladder carcinoma is found in 1-2% of patients who undergo surgery on the biliary tract. Although the exact etiology of carcinoma of the gallbladder is unknown, cholesterol gallstones are commonly implicated by association[3]. Most patients suspected of carcinoma of the gallbladder have advanced disease. Discovery of patients with early disease usually is fortuitous, the diagnosis of carcinoma being established only positively after histologic examination of gallbladder removed for cholecystitis. Carcinoma gallbladder has a poor prognosis despite surgical resection of the tumour[4].

## PATIENTS AND METHODS

This prospective study was carried out in the Department of Surgery, Surgical Unit-II, Chandka Teaching Hospital, Larkana, from January 1997 to December 1998. Majority of these patients were admitted through O.P.D and some were referred from medical wards. Data collected on proforma included sex, age group, socio-economic group, mode of admission, presenting symptoms and any associated disease. Relevant findings on general physical and abdominal examination were also recorded.

Necessary laboratory investigations were carried out preoperatively, which included complete blood picture, urine analysis, blood sugar, blood urea, serum electrolytes, L.F.T.s and serum anylase. Imaging techniques, such as X-ray chest, abdominal ultrasonography and P.T.C. (in some of the cases, if ultrasound confirmed the intra and extrahepatic duct dilatation) were used. All the patients were operated as soon as they were investigated. Various procedures were performed, which included cholecystectomy, cholecystectomy with exploration of C.B.D. and T-tube drainage, cholecytectomy with wedge excision of liver, biopsy of the mass or lymph node. Thorough examination of gallbladder, extrahepatic biliary tree, cystic lymph node, liver, spleen, pancreas, omentum and duodenum was carried at the time of surgery. Gross examination of removed gallbladder alongwith its interior was performed in the operation theatre. Gallbladder and any other tissue for biopsy were preserved in formalin and sent for histopathological examination. Removed gallstones were also sent for chemical analysis.

Correspondence:
Dr. Sher Mohammad Shaikh
Department of Surgery
Chandka Medical College, Larkana

## RESULT

During the study period of two years, we examined hundred patients (80 female and 20 male) with cholelithiasis and ten of them had carcinoma gallbladder. Six (60%) were females and 4 (40%) were males (female to male ratio 1.5.1). Their ages ranged from 41 to 68 years with mean age of 52.2 years, five out of the ten patients were in the age group of 40-49 years, three were in the age group of 50-59 years and two were in the age group of 60-69 years. Carcinoma gallbladder was more common in the 5th decade of life. Majority of the patients, (nine out of ten) belonged to poor families.

Common presenting symptoms were abdominal pain (100%) nausea and vomiting (80%) weight loss (40%) jaundice and anorexia (20%). Abdominal mass was palpable in two. Puritis fever was noted in each patient (Table-I).

**TABLE-I FREQUENCY OF SYMPTOMS AND SIGN**

| Symptoms | No of Patients | % |
|---|---|---|
| Pain | 10 | 100 |
| Nausea & vomiting | 8 | 80 |
| Anorexia | 2 | 20 |
| Weight loss | 4 | 40 |
| Jaundice | 4 | 40 |
| Pruritis | 3 | 40 |
| Fever | 3 | 30 |
| Mass abdomen | 2 | 20 |

Preoperative diagnosis with the help of ultrasound was possible only in three patients. Patients with signs of tumour outside the biliary tree were diagnosed positively. This suggests that ultrasound was only helpful in diagnosis of advanced stage disease.

On histopathology, adenocarcinoma was the type of tumour in all the cases. In two patients growth was well differentiated, in five moderately differentiated and in three it was poorly differentiated. Gallstones could not be removed in two cases because of inoperability of the tumour. However, the gallstones detected by ultrasound were roughly 3-4 cms in size and solitary. In eight cases stones were removed, seven (87.5%) had mixed stones with cholesterol as a predominant component, only one had pigment stones.

## DISCUSSION

The lack of specific signs and symptoms at an early and resectable stage delays diagnosis of carcinoma of gallbladder[2]. It is very difficult to diagnosis preoperatively and is often unexpectedly discovered during surgery for benign gallbladder disease. However, when it present clinically, it has already advanced to an inoperable stage.

Review of literature shows a high association of carcinoma of the gallbladder with cholelithiasis. Although the exact cause of carcinoma gallbladder is unknown, the most commonly implicated condition is cholesterol cholelithiasis[3,5]. The overall incidence of carcinoma of the gallbladder in patients with cholelithiasis has been reported 2.7% by Riaz and Ali[6], 6.6% by Shah et al[7] and 6.1% by Rodriguez-Obero et al[8]. In our study, the incidence of carcinoma gallbladder with cholelithiasis is 10%, which is higher than in other studies. The higher figure in our study might be due to late presentation of patients with cholelithiasis, as it is not unusual that many years elapse before their first symptoms appear. Either the patients ignore the symptoms or they are misdiagnosed as acid peptic disease. The prolonged irritation caused by gallstones may lead to metaplastics, dysplastics and malignant changes [9,10,11,12,13]. Out of eight patients in whom gallstones could be removed and analyzed chemically, it was noted that in 87.5% of the cases the stones were mixed with cholesterol as a predominant component.

In our study, 60% patients of carcinoma of the gallbladder with cholelithisis were females and 40% males with female to make ratio of 1.5.1. Results are more or less consistent with other local and international series. However, male patients presenting with cholelithisis are more prone to have carcinoma of gallbladder[14]. In our study carcinoma of gallbladder occurred in 20% of the male patients and 7.5% of the female patients with cholelithisis. Male to female ratio ranges from 1:1.4 to 1.4 [2,6,15,16,17,18]. Higher prevalence of carcinoma of gallbladder in male patients with cholelithisis has also been reported by Riaz & Ali, which shows that among male patients it was 9% and among females it was 2%.

Nadler & Mesberry[19] have mentioned that carcinoma of the gallbladder is a disease of elderly female patients with cholelithisis. In our study 66.6% of male patients were in the sixth decade of life and 80% of female patients were in the fifth decade of life. These results are in accordance with those of Riaz & Ali[6]. Pichler & Crioblow[2] have reported mean age of patients with carcinoma gallbladder as 66.2 years. Parakevopoulos[17] has reported mean age of 76 years, minimum age of 49years and maximum age of 90 years. Mean age of patients in our study is 52.2 years, with minimum age of 41 years and maximum age of 68 years. This is much nearer to that described by Riaz & Ali[6], but lower than that described by Henson et al[18].

The presentation of an early curable carcinoma of gallbladder is usually identical to that of benign gallbladder disease[1]. Pain is the most prevalent symptom as described by Wanebo in 79%. & Koo et al in 81% cases[3,20]. Nausea and vomiting are less common in studies by Wanebo (50%) & Koo (8%). Incidence of weight loss is similar to that described by Wanebo (42%) and Koo J (50%).

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Mass abdomen was found in 20% patients of carcinoma of gallbladder in our study. This finding is similar to that described by Houry[21] (15%) and Paraskevopoulos[17] (14.29%) but Wanebo[20] has shown that mass abdomen was present in 29% cases. Adenocarcinoma was the most prevalent histologic type of gallbladder carcinoma in this study. Other studies have shown 75-80% prevalence of carcinoma the gallbladder with cholelithisis[6,16,22,23].

In our study, growth was well differentiated in 20% cases moderately differentiated in 50% and poorly differentiated in 30% which is consistent with some other studies[8].

Preoperative diagnosis and prediction of the extent of disease could be advantageous for two reasons. First, the patient and Surgeon would be prepared for major curative resection, if possible. Second when the disease has progressed too far, operation could be postponed until essential for pallation[24]. Ultrasonography seems to be the most appropriate tool for detecting early carcinoma of gallbladder[1].

Preoperative diagnosis of this disease with the help of ultrasonography was possible only in 30% patients, with advanced stage disease. This result is in accordance with that of Riaz & Ali[6], who could only diagnose 24% cases. However, preoperative diagnosis was much higher than that described by Parakevopoulos[17], who could diagnose only in 9.5% cases and is much lower than that by Sovia [23] who reported preoperative diagnosis in 75% patients. Carcinoma of gallbladder has a poor prognosis despite surgical resection[4]. There is no statistically significant difference in survival after extended or simple cholecystectomy in stage-I disease[26].

Radiation therapy has been used in an attempt to improve the prognosis, but only a few series of less than ten cases have been reported. Radiotherapy may increase survival after no resection or palliative resection of gallbladder carcinoma[21]. There was no difference in the survival between patients treated or not treated with chemotherapy.

## REFERENCES


1. Koga A. Yamauchi S, Izami Y and Hamanka N Ultrasonographic detection of early and curable carcinoma of the gallbladder. Br. Jr. Surg. 1985, vol. 72-728-30.
2. Pichler JM Crichlow Rw. Primary carcinoma of the gallbladder. Surg. Gynecol Obstet. 1978; 147:929-42.
3. Koo J. Wong J. Frank C.Y. Cheng, Ong B Carcinoma of the gallbladder. Br. J. Surg, 1981; 68:161-65.
4. Metha A, BahadurA.K, Aranya R.C. Jain A.K. Role of radiation therapy in carcinoma of the gallbladder – a preliminary Indian experience Trop. Gastroenterol 1996; 17:22-25.
5. Hart. J. Modan B, Shani M. Cholelithisis in the etiology of gallbladder neoplasm Lancet 1971; 1:1151-53.
6. Riaz S. Ali A. Carcinoma of gallbladder. Pak J. Med. Res. 1989; 28:267-269.
7. Shah S.A., Shami I.H., Bhaty M.A. Gall stones disease in Pakistan. The J Surg. 1990; 1:30-33.
8. Rodriguez-Otero J.C., Rainoldi J, Celoria G, Monti J. Acute cholelithisis and cancer of the gallbladder in the aged Acta Gastroenterol Latinoam 1985; 15 (2): 123-5.
9. Lowenfels A.B., Lindstorm C.G., Conway M.J., Hastings P.R. Gall stones and risk of gallbladder cancer J. Natl. cancer Inst. 1985; 75(1): 77-80.
10. Hisatomi K, Haratake J, Horie A, Ohsato K. Relation of histopathological features to prognosis of gallbladder cancer. Ann J. Gastroenterol. 1990; 85(5): 567-72.
11. Cotran R.S, Kumar V., & Robbins S.L. Robbins pathologic basis of disease 4th edition W.B. Saunders International edition. 1989,974-5.
12. Yangagiva H, Onishi N., Tenada. The relation of stone and carcinoma of gallbladder based on mucosal findings Gan. No. Rishno. 1990; 36(12):2131-36.
13. Yamato M., Nakajo S. Tahara E. Gall stones in gallbladder diseases. Acta Pathol. Jpn. 1989; 39(9): 582-5.
14. Maringhimi A, Morean J.A., Melton L.J. Hench V.S., Zinsmeister A.R. Dimango E.P. Gall stones gallbladder cancer, and other gastrointestinal malignances. An epidemiological study in Rochester, Minnesota Ann Inten. Med. 1987; 107(1): 30-5.
15. Nervi F., Duratel, Gomez G. Rodriguez G, Del Pinog, Ferreiro G. Frequency of gallbladder cancer in Chile, A high risk area. Int. J. Cancer 1988,41(5): 657-60.
16. Henson DE., Albores-Savedra J., Corie of carcinoma of the gallbladder. Histologic types stages of disease, grade & survival rates. Cancer 1992,70:1993-97.
17. Paraskevopoulos JA, Dennison AR, Ross B. Johnson. A.G. Primary carcinoma of the gallbladder: a 10 year experiences Ann R. Coll. Engl. 1992; 74(3): 422-24.
18. Moreaux J, Dachy C. Cancer of the gallbladder. A surgical experience. Presse Med. 1989; 18(37): 1836-39.
19. Nadler L.H., McSherry C.K. Carcinoma of the gallbladder: review of the literature & report on 56 cases at the Beth Israel MedicL Center. Mt. Sinai J. Med: 1992, 59(1): 47-52.
20. Wanebo H.J. Carcinoma of the gallbladder. In Wanebo H.J. (ed) Hepatic & Biliary cancer. New York Marcel Dekker 1987;432.
21. Houry S, Schlanger m, Hugnier M, Lacaine F., & Langier a Gallbladder carcinoma role of radiotherapy. Br. J. Surg 1989; 76:448-450.
22. Renard P., Falvre J. Boutran M.C., Milanc Bedenne L, Klepping C. Cancer of the gallbladder in the cote d or Gastroenterol Clin. Biol. 1987; 11(3):237-41.
23. Sons H.U. Borchard F., Joel B.S. Carcinoma of the gallbladder. Autopsy findings in 287 cases & review of the literature J. Surg. Oncol. 1985; 28(3):199-206.
24. Moos A.R., Hall A.W., Moraldi A. Skinne D.B. The continuing challenge of gallbladder cancer. Ann J. Surg. 1975; 130:57-62.
25. Sovia M, Aro K. Panula M. et al. Ultrasonography in carcinoma of the gallbladder. Acta Radiol. 1987,28:711
26. Kapoor V.K., Haribhakti S.P. Extended cholecystectomy for carcinoma of the gallbladder. Trop. Gastroenterol. 1997; 16(4) 74-75.

# MODIFIED BLALOCK-TAUSSING SHUNT: A PALLIATIVE PROCEDURE FOR CONGENITAL CYANOTIC HEART DISEASE

ABID A. AWAN, IQBAL A.K., MALIK A., M. REHMAN.

*ABSTRACT:*
*Systemic pulmonary arterial shunt procedure is used in the management of cyanotic congenital heart diseases. We are reporting our experience of 80 procedures with modified Blalock-Taussing shunt procedure in the management of cyanotic heart defects, from January 1994 to December 1994. The age of these patients was 3 months to 15 years. Forty-five (56.2%) presented with cyanotic spells and 35 (43.5%) were electively selected for surgery. The packed cell volume in majority of the patients ranged between 45 to 70%. Seventy-two (90.5%) cases received right-sided shunt. The hospital mortality was 5%. We conclude modified Blalock Taussing shunt is a useful palliative procedure in the management of cyanotic congenital cardiac defects.*

*KEY WORDS: Congenital heart diseases, modified Blalock-Taussing shunt.*

## INTRODUCTION

Systemic-pulmonary artery shunts continue to play an important role in the management of cyanotic congenital cardiac defects[1], either as a palliative procedure prior to total correction, or as the sole means of palliation for defects not reparable at the present time[2]. Since the introduction of a subclavian pulmonary artery shunt by Blalock and Taussing in 1945, a number of systemic-pulmonary artery anastomosis have been described. We present our experience with modified Blalock-Taussing shunt procedure in the palliation of cyanotic heart defects.

## MATERIAL AND RESULTS

From January 1994 to December 1994, 80 modified Blalock-Taussing shunts were carried out. Age range was 3 months to 15 years. There were 44 (55%) male and 36 (45%) female patients. The distribution of patients according to their diagnoses is shown in Table-I.

In all the cases the indication for surgery was hypoxemia caused by partially or completely obstructed pulmonary blood flow, 45 (56.2%) patients presented with cyanotic spells, while 35 (43.5%) were electively selected for surgery. Their packed cell volume (PCV) range is shown in Table-II.

**TABLE-I DIAGNOSIS OF PATIENTS**

| Diagnosis | Number | % |
|---|---|---|
| Tetrology of Fallot | 44 | 48.7 |
| Tetrology of Fallot with Pulmonary Atresia | 5 | 6.2 |
| Transposition of Great arteries | 18 | 18.2 |
| Tricuspid Atresia with Pulmonary steonsis | 5 | 6.2 |
| Single Venticle with Pulmonary Atresia | 3 | 3.2 |
| Double Outlet right Ventricle with Pulmonary steonsis | 5 | 6.2 |

**TABLE-II PCV RANGE**

| PCV Range | Number | % |
|---|---|---|
| Less than 45% | 14 | 18 |
| 45-60% | 42 | 52 |
| 60-70% | 18 | 22 |
| 70-80% | 4 | 8 |

Modified Blalock-Taussing shunt (B-T Shunt) was carried out in all the cases. Seventy-two (90.5%) cases received right-sided shunt, while only 10% of the cases received left B-T shunts. The chest was opened through a lateral thoracotomy through fourth intercostal space. The size of the graft was related to the existing anatomy. Normally the

Correspondence:
Dr. Abid Aslam Awan,
Department of Cardiac Surgery,
National Institute of Cardiovascular Diseases Karachi.

largest diameter graft that could be conveniently anastomosed to the subclavian and pulmonary artery was chosen 6-7mm grafts was used in majority of cases. Both ends were anastomosed using continuous 6/0 prolene. Patients were anticoagulated with heparin (1mg/kg). Our hospital mortality figure were 5%, as shown in Table III.

**TABLE-III HOSPITAL MORTALITY**

| Diagnosis | Age | Weight | Cause |
|---|---|---|---|
| Transposition of great arteries | 2 months | 2.3 kg | Arrhythmia (Intraoperative) |
| Single ventricle with Pulmonary atresia | 3 months | 2.2 kg | Hypothemia Bradycardia |
| Tetrology of Fallot with Pulmonary artresia | 4 months | 3.0 kg | Metabolic Acidosis |
| Tricuspid atresia with Pulmonary steonsis | 6 months | 4.3 kg | Metabolic Acidosis |

One of the patients died intraoperatively due to arrhythmia. Three of the cases died in the intensive care unit secondary to metabolic acidosis and arrhythmias. All the surviving cases had adequately effective palliation. Their arterial oxygen saturation improved. The early non-fatal complications that we came across are shown in Table-IV.

**TABLE-IV EARLY NON-FATAL COMPLICATIONS**

| | |
|---|---|
| Pulmonary edema | 3 |
| Chylothorax | 1 |
| Inotrope Support | 3 |
| Chest wall bleeding | 1 |
| Serous effusion | 2 |

Three patients with pulmonary edema were treated with diuretics and were kept on ventilation for an average of 24 hours. One of the patient developed chylothorax which did not respond to conservative measures. Exploratory thoroacotomy was done and thoracic duct was ligated. Two patients developed moderate serous effusion within a week after surgery and were treated with pleural aspiration.

## DISCUSSION

The value of systemic-pulmonary arterial shunt procedures as palliative treatment for cyanotic patients with diminished pulmonary blood flow secondary to congenital heart diseases has been well established.[3] Despite recent advance in the repair of congenital heart disease in infancy[4], systemic-pulmonary artery shunts continue to play an important role in the management of such defects[1]. There are three basic aims of palliative shunt surgery: to improve the pulmonary blood flow, to allow growth of the pulmonary vasculature and symptomatic improvement. Though definitive surgery is being offered to neonates and infants with cyanotic heart disease, palliative surgery still has certain definite indications:

- Lack of facilities for total correction of congenital cardiac lesion.
- Severely cyanotic sick baby.
- Abnormal pulmonary vasculature like pulmonary artery hypoplasia, pulmonary artery branch stenosis, nonconfluence of pulmonary arteries, pulmonary artery arbolization anomalies.
- Descending coronary artery crossing the right ventricular outflow tract, which mandates the use of an extra cardiac conduit to achieve correction and multiple ventricular septal defects.

Modified Blalock-Taussing shunt is much closer to an ideal shunt, as it is easy to construct, provides effective pulmonary blood flow, allows growth of the pulmonary vasculature, does not distort pulmonary artery, does not lead to pulmonary hypertension, has reliable early and late flow characteristics, is comparatively easier to take down at the time of definitive surgery and has low operative risk[6].

Disadvantages of Blalock-Taussing shunt are neo-initial proliferation leading to graft occlusion, infection of the prosthetic material, serious leakage and seroma formation[7]. False aneurysm is a rare complication of modified Blalock-Taussing shunt. It may present with fatal-hemoptysis[8]. Fibrous reaction around the graft leads to adhesions[9] and difficult dissections at the time of definitive surgery with increased risk of post operative mediastinal bleeding.

## REFERENCES


1. De Leval RM, Mckay R. Jones M et al. Modified Blalock-Taussing shunt: use of subclavian artery orifice as flow regulator in prosthetic systemic pulmonary artery shunts. J Thorac Cardiovas Surg 1981; 81:112-119.
2. Arcinegas E. Farooki QZ, Hakimi M et al, Classic shunting operating operations for congenital cyanotic heart defects. J. Thorac Cardiovas Surg, 1982;84:88-96.
3. Truccone JN, Bownan of, Malm RI, and Gersony MW. Systemic pulmonary arterial shunts in the first year of Life. Vol. 49.
4. Eastaneda A., Fareed M, William SR. Norwood W. Repair of tetrology of Fallot in infancy. Early and late results. J. Thorac Cardiovas Surg, 1979;74:372-381.
5. Stephenson LW, Frieman S, and Edmunds LH: Staged Surgical management of Tetrology of Fallot in infants. Circulation, 58; 837-841:1978.
6. Lamberti J, Carlisle J, Waldman JD, Lodge FA and Kirkpatrick Se et al. Systemic–pulmonary shunts in infants and children–J. Thorac Cardiovas Surg, 88; 76-81: 1984.
7. Bove EL, Kohman L, Sereika S, Bym CJ, Kavey RW, Blackman MS, Sondheimer HM, Rosenthal A: The modified Blalock-Taussing Shunt analysis of adequacy and duration of palliation–Circulation 1987:76 (Suppl. III: 19).
8. Sethia B, Pollock JCS–False aneurysm formation: A complication following the modified Blalock-Taussing shunt. Ann Thorac Surg. 1986: 41:667.
9. Kay H, Caupuani AD, Frankes R, Lincoln C, Experience with the modified Blalock-Taussing Operation using polytetraflouroethylene grafts. Br. Beart J 1983; 49:359-63.

# DAY CARE SURGERY FOR CLEFT LIP IN ADULTS

M.A. NAUSHAD BEG

*ABSTRACT:*
*Cleft lip repair by Millard's technique was done on 13 patients, all males, at day care surgery in Chandka Medical College Hospital Larkana. Their ages were between 20 to 35 years. Local anaesthesia used was 1% Lignocaine with Adrenaline. The results were excellent with no untoward side effects; there were no complications.*

*KEY WORDS: Cleft lip, Surgery*

## INTRODUCTION

Repair of cleft lip dates back to 390 A.D. in China. However, the first documentation was by Flemish surgeon Yperman (1295-1351). In 1556, Franco of Switzerland first used the term "cleft lip" instead of hare lip and fully described its repair, using needles, adhesive stents and relaxing incisions. In 1597, Tagliocrozzi made use of mattress sutures. Pioneer cleft lip surgeons include Yperman, Franco, Von Graefe, Veau, Rose and Thomson were doing straight-line repairs. After a lapse of 250 years Mirault in 1855 started using flaps. Gustav Simon of Heidelberg started using a medically based square flap in 1864. Hagedorn reversed the Simon repair and Le Mesurier further modified it with remarkable results. In 1952 Tannison saved the cupids and it worked very well, marking beginning of the modern era. All repairs today save the cupids bow on the cleft side of midline and create a medial defect, filling it with lateral tissue. In a major development in cleft lip repair, Millard realised that cupid bow was displaced as a unit and needed only downward rotation. He suggested that the scar should fall in the line of the philtral ridge. Repair created a medial defect by a single curved incision, followed by a direct lateral to medial advancement. Skoog of Sweden further worked on Milliard's repair to help correct the alar distortion.

## PATIENTS AND METHODS

Cleft lip repair was carried out on 13 patients, all adult males, between the ages of 20 and 35 years. All the cases were done under local anaesthesia; Lignodcaine 2% with adrenaline, 1:200,000 diluted to half its strength, with the addition of equal amount of distilled water, was infiltrated into the surrounding tissues. Not more than 20 ml of the local anaesthesia was used in a case.

Twelve cases were of unilateral cleft lip with no cleft palate, 8 were right sided. A single case of bilateral cleft lip with associated complete cleft of palate was also done under local anaesthesia touching only the cleft lip. Patients were called on the day of surgery with no specific instructions, except shaving their moustaches before coming.

A proper history to exclude any associated congenital deformity, bleeding disorders and any drug allergy was particularly taken. No pre-operative investigations were done. Millards technique of rotation with medial advancement flap was done in 12 cases of unilateral cleft lip. There was a single case of bilateral cleft lip where a simple repair of both sides was done in one stage. Orbicularis muscle contracture release was accomplished as a unit with the overlying skin. Separate muscle dissections were not used. Chromic catgut 4-0 sutures were used for deeper layers and 4-0 silk was used for the skin. The skin stitches were removed on the 5th post-operative day.

After the operation, the patients were sent home with no dressing except a thin film of antiseptic ointment. They were advised to be on fluid diet for the first two days using a straw to sip the fluid. Oral antibiotics and analgesics were given with instructions to return on the fifth post operative day for removal of stitches.

Correspondence:
Dr. M. A. Naushad Beg,
Assistant Professor of Surgery, D.M.C. / Civil Hospital, Karachi.

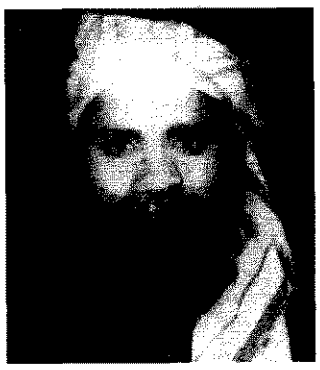

**Figure 1 (a)**
Pre-operative defect

**Figure 1 (b)**
Post operative result

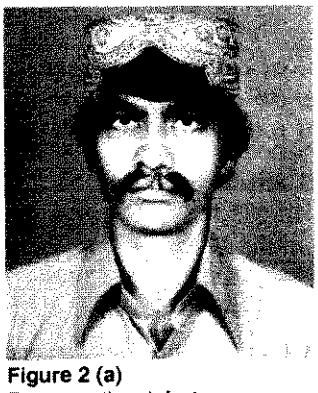

**Figure 2 (a)**
Pre-operative defect

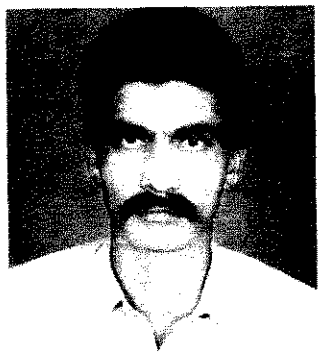

**Figure 2 (b)**
Post-operative result

## RESULTS

The results were excellent as all the patients were adults with no need of general anaesthesia and least risk of secondary growth distortion. There were no cases of wound infection or wound disruption, with zero mortality. However in the single case of bilateral cleft clip, the patient returned the same night after the operation with oedema of the lips and cheeks. Patient was admitted for the night just for observation and discharged in the morning as the oedema subsided without any specific medication. Thereafter the wound healed satisfactorally.

## DISCUSSION

As pointed out by Millard, the "Rule of 10" should be observed in cleft lip repair: 10 1b weight, 10 Gm haemoglobin and 10 weeks of age. However, surgery can be performed with greater safety and with least risks of secondary growth distortion if the patient is an adult. In remote places in the province of Sindh, where education is at the lowest ebb and people are not much concerned about their looks, it was found that most of the cleft lips, attained their adulthood without any social difficulty. In fact, it was found that most of these patients were married. However, this is not always the case; at least two young men came with cleft lip saying that their fiancees have refused to marry them unless their facial defects were corrected by surgery. Also a case of bilateral cleft lip, working as a waiter in a roadside canteen, was told that he might lose his job as he was scaring away the customers. (Figures 1 & 2)

## REFERENCES


1. Boo Chim K. An ancient Chinese text on cleft lip. Plast Reconstr. Surg 98 PP 189.
2. McCarthy JG, Cutting CB Hogan VM in: McCathy JG (Ed) Plast Surgery (Vol. 4, Philadelphia, WB, Saunders 1990 PP 2437-2450.
3. Byad H.S. Plastic Surgery 4th ed. 1991 (Chap 9 & 10) 271-285.
4. Ravitch – JA, William K. Welch KJ: et al Paediatric Surgery. 4th ed. Ch. 47. Chicago Yen book medical publishers Inc. 1986 PP 451-460.
5. Bloom Herbert, et al Surgical Repair of cleft lip and mod Maxillofacial Surgery Vol. II by W. Harry Archer. 5th ed. 1975 W.B. Saunders Company Ch. 30:1841-1848.

# AN EXPERIENCE WITH POLYPOID DISEASE OF GASTROINTESTINAL TRACT

DAULAT KHAN

*ABSTRACT:*
*Polyps are relatively common in infants and children. Most of them are benign lesions and a few are associated with syndromes. In a year's period from August 1998 to July 1999, 120 patients with gastro intestinal polyps were managed at the Sandeman Provincial Hospital, Quetta. There were 68 male patients and the age ranged from 3 years to 13 years. All patients with colonic polyps presented with rectal bleeding while poly prolapse and rectal prolapse were major complaints in 10 and 5 patients respectively. The histology in all these were of juvenile type. One 13 years old female patient presented with acute abdominal pain and vomiting. She had café au lait spots. She was explored in emergency and found to have ileo-ileal intussusception due to polyps. Resection and anastomosis was performed. The biopsy revealed Peutz Jeghers syndrome.*

*KEY WORDS: Juvenile polyp, Peutz Jeghers syndrome.*

## INTRODUCTION

Polypoid diseases of the gastrointestinal tract include isolated polyps, uncommon syndromes and as a manifestation of disorders in other parts of the body[1]. Polyps are common cause of rectal bleeding in age group of 3-5 years[2]. Most of the polypoid lesions are non neoplastic, hamartomatous malformations but malignant lesions also occur[3]. In this study our experience of gastrointestinal polypoid lesions is described.

## PATIENTS AND METHODS

This is a one year retrospective review of the record of all the 120 patients who were managed at Sandeman Provincial Hospital, with the diagnosis of polypoid lesions of gastrointestinal tract during the period of August-98 to July-99. The data collected include age, sex, clinical presentations, management and review of histopathology reports. In all the patients per rectal examination was performed in out-patient department. The patients in whom polyp was felt were given appointment for elective polypectomy after bowel preparation. All procedures were performed under general anaesthesia. The standard technique of polypectomy was employed in all cases. All specimens were sent for biopsy.

## RESULTS

In a year's period 120 patients were managed. There was slight male predomination (male 68, female 52). The age ranged from 3 to 13 years but most of the patients (84) were less than 6 years of age. Rectal bleeding was the main complaint in all the cases. Bleeding was in the form of fresh drops at the time of daefecation. In ten patients mothers noticed polypoid lesion at the anal verge (Fig-1), while five patients had rectal prolapse as well. Single polyps were removed in all the cases. The biopsy report of 119 patients revealed juvenile polyp.

One 13 years old female patient presented with acute abdominal pain and vomiting. There was history of maelena in the past with repeated episodes of abdominal pain. On examination *café au lait* spots were seen (Fig-2). Abdominal X-ray showed pattern of small bowel obstruction. The provisional diagnosis of Peutz Jeghers syndrome with intussusception was made. Following correction of fluid and electrolyte imbalance patient was operated. At laparatomy ileo-ileal intussusception was found. The segment of the gut contained large polyps, which were the cause of intussusception. This segment was resected and anastomosis was performed. Many small polyps were also present in the bowel. The post operative recovery was uneventful. Biopsy report revealed hamartomatous polyps.

## DISCUSSION

Juvenile polyps were first described by Verse in 1908 although the term juvenile polyp was first used by Horrilleno in 1957. It was Morson (1962) who was the first to

Correspondence:
Dr. Daulat Khan,
Assistant Professor, Department of Paediatric Surgery
Sandeman Provincial Hospital, Quetta

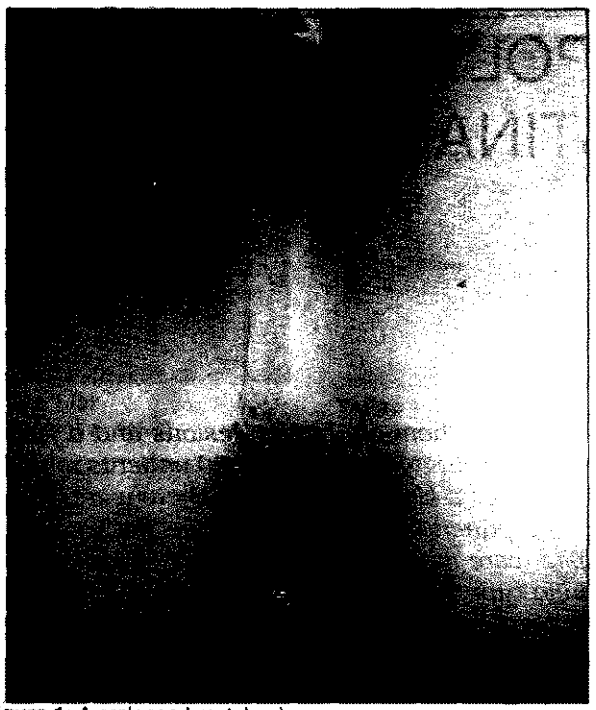

**Figure 1:** A prolapsed rectal polyp

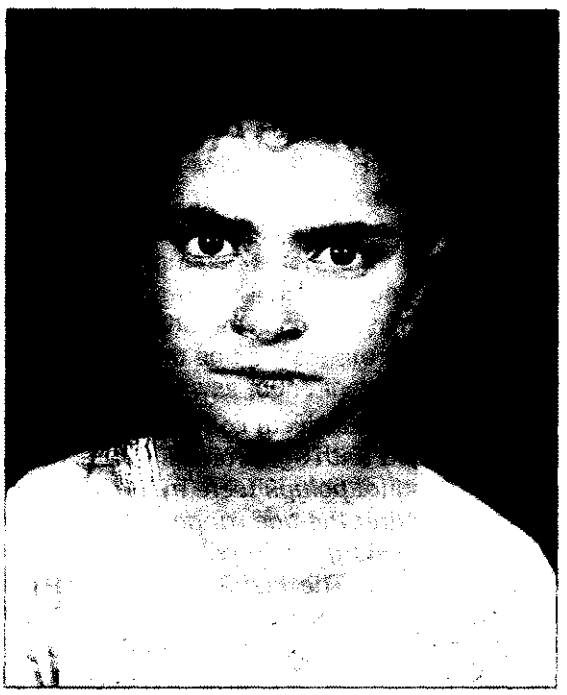

**Figure 2:** A patient of Peutz Jeghers syndrome with *Cafe au lait* spots on face.

demonstrate that these polyps were benign hamartomatous lesions. Before that all the polyps were considered as adenomas[4]. Juvenile polyps are also called inflammatory or retention polyps. They are the common cause of rectal bleeding in children. Usually they are isolated lesions but multiple polyposis with and without associated syndromes are also described[5]. Most of them are found in the rectum and are usually solitary. The diagnosis is clinical, easily made on rectal examination. Simple procedure of polypectomy rids the patient of the annoying symptoms.

Peutz Jeghers syndrome is the association of intestinal polyps with mucocutaneous pigmentation and was first reported in 1921. The spots are melanotic in nature, ranging in colour from brown to black, occurring on the lips, around the mouth and buccal mucosa[6]. They can be found in other regions of the body as well. The polyps can be found anywhere in GIT but in more than 55% of cases are seen in small bowel[7]. The polyps range from few millimeters to several centimeters in size. Histologically they are also classified as hamartomatous lesions[8]. The relatives of patients should also be screened for such lesions as in most of the cases, autosomal dominant inheritance is found[9]. The association of malignancy, both intestinal and extra intestinal, is high in this condition that demands long term followup of these patients[10].

Our study is in conformity with those reported in literature. Juvenile rectal polyp was the commonest lesion found. Bleeding per rectum in any child demands gentle per rectal examination, which will reveal the polyp if present, in almost all the cases. Invasive investigations are usually not required. Polypectomy, a simple procedure performed as a day care surgery on a prepared bowel under general anaesthesia, is not found to be associated with any morbidity.

## REFERENCES


1. Lelli JR, Coran AG. Polypoid disease of the gastrointestinal tract. In O'Neill JA, Rowe MI, Grosfeld JL, Fonkalsrud EW, Coran AG. (Ed.) Pediatric Surgery, Fifth Edition. Ch. 86, Mosby – Year Book Inc, 1998. 1283 –1296.
2. Mestre JR. The changing pattern of juvenile polyps. Am J Gastroenterol. 1986. 81, 312.
3. Coburn MC et al. malignant potential in intestinal juvenile polyposis syndromes. Ann Surg Oncol. 1995. 2, 386.
4. Morson BC. Some peculiarities in the histology of intestinal polyps. Dis Colon Rectum. 1962. 5,337.
5. Sachatello CR. Gastrointestinal polypoid disease. J Ky Med Assoc. 1974. 75, 540.
6. Phillipis RKS, Spigelman AD. Peutz Jeghers syndrome. In Phillips RKS et al (Ed.). Familial adenomatous polyposis and other polyposis syndromes. Little Brown, Boston. 1994.
7. Tovar JA, Eizaguirre AA, Jimenez J. Peutz Jeghers syndrome in children: Report of two cases and review of literature. J Paediatr Surg. 1983. 18, 1.
8. Soper RT. Gastrointestinal neoplasms. In Ashcraft KW, Holder TM (Ed.) Pediatric Surgery, Second Edition, Ch. 38. W. B. Saunders company. 1993. 453-469.
9. Giardiello FM et al. Increased risk of cancer in Peutz Jeghers syndrome. N Eng J Med. 1987. 1157.
10. Cochet B et al. Peutz Jeghers syndrome associated with gastrointestinal carcinoma. Report of two cases in a family. Gut. 1979. 20, 169.

# AN ANALYSIS OF 30 RENAL BIOPSIES IN PATIENTS SUFFERING FROM NEPHROTIC SYNDROME

BILQUIS LAKHANY, KHAIM CHAND N. MOORANI

*ABSTRACT:*
*This study was conducted at the National Institute of Child Health, Karachi from January 1994 to December 1998 to look for the histological diagnosis of children presenting with nephrotic syndrome, who were either steroid dependent or steroid resistant. Total number of cases were 30 with 20 males and 10 females. Maximum numbers of patients were between 5-10 years of age. Minimal change nephrotic syndrome was the most common diagnosis accounting for 70.37% of cases. Membrano-proliferative glomerulonephritis ranked second (18.18%) followed by focal segmental glomerulosclerosis (11.11%). There is a need to develop better diagnostic facilities for these cases including immunoflourescence studies and/or electron microscopy.*

*KEY WORDS: Renal biopsy, Nephrotic syndrome, Diagnosis.*

## INTRODUCTION

Renal diseases account for approximately 5% of the hospital admissions in a general Paediatric unit[1]. Amongst these, Nephrotic Syndrome is the most common disease. Majority of the children have minimal change disease, however 10-15% may have other than minimal change disease such as membrano-proliferative glomerulonephritis or focal segmental glomerulosclerosis. Most children achieve a complete remission when treated with steroids, though most responsive cases of nephrotic syndrome are likely to relapse. Some children who respond to oral steroid therapy can only be maintained in intermission with relatively high doses of prednisolone. These children are called steroid dependent nephrotic syndrome. In addition one group of nephrotic children who fail to go into remission when treated with oral prednisolone are called steroid resistant. Renal biopsy is done in underlying histopathology[2].

## PATIENTS AND METHODS

A retrospective review was conducted of all the children who underwent percutaneous renal biopsy between January 1994 to December 1998. All biopsies were performed by a radiologist under ultrasound imaging using a trucut disposable needle. No major complication occurred. Indications for doing a renal biopsy were steroid resistant nephrotic syndrome, steroid dependent nephrotic syndrome. In follow up cases laboratory test was performed, which included haemoglobin, haematocrit, prothrombin time and platelet count. Blood pressure and pulse were monitored during and after the procedure. Serial samples of urine, (post biopsy), were examined for haematuria.

All patients received intravenous sedation with 0.1 mg/kg to 0.3 mg/kg diazapam. Local analgesia with 1% lignocaine was used effectively. Pulse and blood pressure were monitored initially at 15 minutes intervals and then less frequently when stable. All urine voided for the next 24 hours was inspected for gross haematuria. Liberal fluid intake was encouraged to promote diuresis and bed rest was advocated for at least 12 hours.

Tissue obtained on biopsy was put in formaline and sent to the laboratory at NICH and immediately, fixed and stained with haemotoxylein and eosin. Total number of glomeruli varied from seven to sixteen. The glomeruli, which appeared as pink dots under light microscopic examination, were counted.

## RESULTS

Biopsies were performed in 30 patients, 20 males and 10 females. Steroid resistant and steroid dependent nephrotic syndromes were the most frequent indications for percutaneous renal biopsy. Twenty patients were between 5 and 10 years of age (Table-I).

Correspondence:
Dr. Bilquis Lakhany,
Assistant Professor,
National Institute of Child Health Karachi.

**TABLE-I** Age & sex distribution

| Age | Male | Female | Total |
|---|---|---|---|
| 2-5 years | 3 | 2 | 05 |
| 5-10 years | 12 | 8 | 20 |
| 10-14 years | 3 | 2 | 05 |
| Total | 18 | 12 | 30 |

Adequate renal tissue was obtained in 27 of the 30 biopsies, giving overall success rate of 90%. Two biopsies contained two to three glomeruli, one contained medullary tissue. Minimal change of nephrotic syndrome was seen in 19 cases (70.37%) while membrano-prolifeative was seen in 5 cases (18.18%) and focal segmental glomerulosclerosis only in 3 cases (11.11%).

## DISCUSSION

Percutaneous renal biopsy is used for diagnosis of renal histology in patients suffering from nephrotic syndrome. Steroid dependent and steroid resistant nephrotic syndrome have been reported to be the commonest indication for renal biopsy in several series [3]. Ultrasound is now widely used as a technique for localization of the kidney. In our series adequate renal tissue for pathological diagnosis was obtained in 90% of biopsies. Edelmann and Grelfer[4] reported a total success rate of 76.5%. Carvajal reported a success rate of 82%. In another large series success rate was 90%. Thus the success rate in the present study is comparable with figures reported by other workers.

In our study minimal nephrotic syndrome was seen in 70.37% cases, which is similar to those described in other parts of the world. However membrano proliferative glomerulonephritis was found on 5 of 30 biopsies giving frequency of 18.18% of all with nephrotic syndrome, which is similar to the study conducted in the Department of Paediatrics, College of Medicine, Ryad, Saudi Arabia[5]. Focal segmental glomerosclerosis was diagnosed on 3 biopsies (11.11%). Clinical complications after renal biopsy includes gross haematuria, passage of clots, severe pain, infections and A.V. fistula. In our study only 9% reported macroscopic haematuria. Nearly half of the parents reported that child had some pain but, no major complication occurred. These results are similar with those reported in literature.

## CONCLUSION

Renal biopsy is an important procedure used to diagnose renal diseases. In small children it is considered a difficult procedure due to the small size and increased mobility of the kidneys. Some Paediatrics centres are still using fluoroscopy and general anaesthesia. We emphasize that intravenous sedation with ultrasound guidance is a safe and reliable method for obtaining an adequate renal specimen without complications.

## REFERENCES

1. Ramzan A. An analysis of renal cases at NICH, Pak Paed. J. 1990 XIV 171-174.
2. Ticher C Croker BP; Indications for renal biopsy: evaluation by light, electron and Immunofluroescence microscopy. In disease of the kidney 4th ed. Little Brown Boston, PP 527-553
3. Edelmann C M. Jr. Chung J. Renal biopsy: Indication, technique and Interpretation in Edelmann C M. JR Paediatric kidney diseases Boston MA. Little Brown. 1992 pp 499-527.
4. Parrish, AE, Complications of percutaneous renal biopsy a review of 57 years experience. Clin Nephrel 38: 135-141.
5. Abdurrahman MB, Elidrissy, Shipkey 4FH, Rashid S, Nouri M, Annual Trop Paediatr, 1990: 19(2): 125-132.

# SCREENING FOR AUTOIMMUNE THYROID DISEASE IN TYPE 1 DIABETICS

SYED IFTIKHAR ALI, JAMAL ARA, RUKHSANA SATTAR, S. SHABEER HUSSAIN.

*ABSTRACT:*
*A prospective study to screen high risk (type 1 diabetes mellitus) subjects for autoimmune thyroid disease, was carried out on 55 patients at JPMC, Karachi from January to December 1998. Thyroid dysfunction was noted in 11 out of 55 patients screened. Microsomal antibody was present in high titre (mean 1:7866.667) in 9 patients and thyroglobulin antibody was found in low titre (mean 1:121.429) in 9 patients. Thyroid dysfunction has significant association with Type-I diabetes. Microsomal antibodies alone are good predictors of autoimmune thyroid disease in these patients.*

*KEY WORDS: Type I diabetes, Autoimmune thyroid disease, Thyroglobulin antibody, Microsomal antibody.*

## INTRODUCTION

Autoimmune thyroid disease is one of the elegant examples of organ specific autoimmune disorders[1]. There is no internationally recognized classification of autoimmune thyroid disease[2]. The spectrum of autoimmune thyroid disease ranges from autoimmune Grave's disease to Hashimoto's disease[3]. Autoimmune thyroid disease can be diagnosed on the basis of clinical suspicion supported by raised titre of anti-thyroid antibodies. These antibodies are directed against thyroglobulin and mircosomal antigens of the thyroid[3]. American Thyroid Association guidelines recommend sceening for autoimmune thyroid disease in susceptible groups of patients[4]. Routine screening of Type-I diabetes is warranted as long as the frequency of thyroid dysfunction is increased in these subjects[5].

## PATIENTS AND METHODS

The inclusion criteria was insulin dependant subject who had been diagnosed as Type-I diabetics using WHO Criteria6. All those patients who were currently not taking insulin or had been diagnosed as Type-II diabetics were excluded. A proforma was completed stating salient features of hypothyroidism and hyperthyroidism. Height and weight of the patients were noted.

Correspondence:
Dr. Syed Iftikhar Ali
Medical Unit I,
Jinnah Postgraduate Medical Centre
Karachi.

Blood samples were obtained from each patient and were immediately submitted for FT3, FT4, and TSH estimation at Atomic Energy Medical Centre, JPMC. Thyroid antibodies samples were maintained in proper cold chain before being assessed.

FT3, FT4 and TSH were assayed using commercially available RIA kits, which measure levels of these hormones by radioimmunoassay, using labeled antibodies bound to particles along with radioactive tracer analogue, which is then measured[7]. Thyroid autoantibodies were measured using haemagglutination tests based on Boyden's principle[8]. For measurement of antibodies commercially available kits[9]. Thymune M and "Thymune T" (Wellcome, Dartford, England) 'U' well type microtitration plates were used. These haemagglutination tests use "Turkey ertythrocytes" which had been coated with human microsomal antigen and human thyroglobulin antigen. These sensitized cells will agglutinate in the presence of specific antibodies. A titre of 1:10 was considered positive for Thymune-T and a titre of 1:100 was considered positive for Thymune-M[8].

Statistical analysis was done using a computer program "Epi Info 6" and a 2x2 table, when thyroid dysfunction was plotted against antibody positivity.

## RESULTS (TABLE 1)

This study included 55 patients between the ages of 12-30 years. The mean age for females was 22.5±6.75 years. The mean duration of type 1 diabetes was

51.65±6.78 months with a mean duration of 48 months in females and 36 months in males. Only 4 out of 25 male patients had goitre and none of them had clinical signs of thyroid dysfunction. Twentyfour out of 30 female patients had goitre. Thyroid dysfunction was diagnosed clinically in 5 patients with diffuse goitre. Among the 55 patients, five (all females) had elevated TSH levels above the cut off value of 4.6 uU/ml and three of these patients had significant titre of microsomal antibodies. $FT^3$ and $FT^4$ were elevated in 2 female patients and this finding correlated well with significant microsomal antibody titre. Sub clinical thyroid dysfunction was noted in 6 patients.

| TABLE-I | RESULTS OF THE STUDY |
|---|---|
| Males | 25 (45.5%) |
| Females | 30 (54.5%) |
| **Age in Years** | |
| • Males | 20 56+- 4.48 |
| • Females | 22 5+- 6.75 |
| **Thyroid dysfunction** | |
| • Clinical | |
| Hypothyroidism | 3 (5.45%) |
| Hyperthyroidism | 2 (3.63%) |
| **Thyroid autoantibodies** | |
| • Microsomal antibody | 9 (16.6%) |
| • Thyroglobulin antibody | 9 (16.6%) |
| • Both | 8 (14.54%) |

Using 2x2 table thyroid dysfunction was plotted against antibody positivity and a sensitivity of 45.5% and a specificity of 86% and a positive predictive value of 50% and a negative predictive value of 95% were obtained.(Table II)

| TABLE-II | | TESTS RESULTS | | |
|---|---|---|---|---|
| | Normal Range | Mean | Standard deviation | P Value |
| FT3 | 2.2-4.71 pg/ml | 3.477 | 2.095 | 0.000 |
| FT4 | 0.85-1.86 ng/dl | 1.499 | 0.709 | 0.000 |
| TSH | 0.5-4.6 | 4.511 | 16.897 | 0.4990 |
| Microsomal antibody | 1: 100 | 7866.667 | 10283.968 | 0.0492 |
| Thyroglobuli n antibody | 1.10 | 121.429 | 166.476 | 0.10020 |

## DISCUSSION

Clinical thyroid dysfunction was noted in 5 of our patients, this is in comparison with a study of McKenna who observed thyroid dysfunction in 7.3% patients9. Clinical hypothyroidism was noted in 3 patients supported by raised TSH levels. Grey noted sub clinical primary thyroid dysfunction in 13% of Type 1 diabetics screened [10].

According to Wickham survey (1977) Thyroglobulin antibody was present in 2% of general population with preponderance among females (3%). Our study showed thyroglobulin antibody to be present in 9 (16.6%) patients. This is comparable to the study conducted by McKenna who has shown thyroglobulin antibody to be present in 7.5% of Type 1 diabetics. In our study microsomal antibody was present in 9 (16.6) patients while McKenna observed microsomal antibody in 15.9% of Type-I diabetics and Wickham survey showed microsomal antibody to be present in 6.8% of general population[11].

An interesting study on Saudi school children in the age group 12-21 revealed microsomal antibody to be present in 7.1% of the subjects studied and was noted in 14.3% of subjects, which was considerably higher using haemagglutination tests in general population; the reason according to authors was consanguinity among parents in that region[12].

Grave's disease was noted in 2 (3.63%) patients in our study compared to a study by Gray who found hyperthyroidism in 3.5% of Type-I diabeties10. Testing is required to measure future thyroid dysfunction and need for further investigation among high risk subjects (family history of autoimmune thyroid disease, elevated titre of microsomal antibody) who require measurement of thyroid dysfunction at 2-3 months interval or earlier[13]. By looking at the results and pattern of thyroid abnormality, we can predict that these patients are liable to develop thyroid abnormality in future and it is important to screen young Type 1 diabetics for thyroid autoimmunity and further studies for thyroid autoantibodies in general population are required in Pakistan

## REFERENCES


1. Roitt, 1: Autoimmune disease; Scope and aetiology in essential Immunology. 8th edition. 1994: pp 383-404; Blackwell Publication.
2. Davies, TF, Amino, N: A new classification of autoimmune thyroid disease. Thyroid, 1993, 4(3), 331-333.
3. Greenspan, FS: The thyroid gland in Basic and clinical Endocrinology, Greenspan FS (editor), 4th edition, 1994, pp 160-226.
4. Singer, PA, Cooper, DS, Leve EG, Ladenson, PW, Braveman, LE, Daniel, S. Greenspan, FS et al: treatment guidelines for patients with hypethyroidism and hypothyroidism. Standards of care committee, American Thyroid Association, JAMA, 1995, 273-808.
5. Mauriceio, D, Detelva, A: Association of diabetes mellitus with autoimmune diseases. Diabetes Rev. Int. Vol. 5 No. 2, April 1996.
6. WHO study group. Diabetes mellitus. WHO technical report series, Geneva WHO, 1987:727.
7. Glover, JS: Monograph III: Labeled antibody methods for the measurement o thyroid hormones in clinical diagnostics. Kodak Thyroid information series 1992.

8. Thymune M and Thymune T: Microsomal and Thyroglobulin antibody kits. Monographs by Wellcome Datford, England. 1994.
9. McKenna, MJ, Herskowitz, R, Wolfsdorf, JI: Screening for thyroid disease in children with IDDM Diab Care, 1990, Vol 13, No. 7, pp 801-803.
10. Gray, RS Borsey, DQ, Seth, J, Herd, R. Brown, NS, Clarke, BF: Prevalence of sub clinical thyroid failure in insulin dependant Diabetes mellitus, J Clin Endocrinol Metab, 1980,50,1034-1037.
11. Tunbridge, WMG, Everest, DC, Hall, R. Appleton, D, Brewis, M, Clarke, F et al: The spectrum of thyroid disease in a community, The Wickham survey Clin Endocrinol, 1977, 481-493.
12. Suleman, RA, Al-Atas, OS El-Desouki, MI, Essa, MA, Al-Nuaim, AA, Al-Sekat, MA, Thyroid antibodies: High prevalence among school children and adolescents in Gizan, Saudi Arabia Med. J. 1995 16 (4). 287-290.
13. Dayan, CM Daniels, GH: Chronic autoimmune thyroiditis, N Eng. J Med, 1996, 335 99-107.

# UNILATERAL ABSENCE OF PULMONARY ARTERY: A RARE CONGENITAL ANOMALY

## A CASE REPORT

ABID ASLAM AWAN

*ABSTRACT:*
*Unilateral absence of pulmonary artery is a rare anomaly. It may occur as an isolated lesion or in association with other congenital cardiac lesions. We present a case of tetrology of Fallots with associated absence of left pulmonary artery in a 25 years old male, with good operative results.*

*KEY WORDS: Pulmonary artery, Congenital heart disease, Tetrology of Fallot, Systemic pulmonary shunt.*

### INTRODUCTION

Unilateral absence of pulmonary artery (UAPA) is a rare anomaly in which either right or left branch of the main pulmonary artery fails to develop[1]. This condition results due to failure of development of proximal sixth aortic arch. UAPA may appear as an isolated anomaly or in association with other congenital cardiac anomalies. We are presenting a case of UAPA associated with tetrology of Fallot (TOF). Pathophysiological aspects of UAPA are discussed.

### CASE REPORT

A 25 years old male patient, who was a diagnosed case of Tetrology of Fallot (TOF) presented with recurrent chest infections and haemoptysis. On examination the patient had pink, congested conjunctiva, central cyanosis and marked clubbing of fingers and toes. Chest was asymmetric with diminished movements on the left side. Grade 4/6 systolic murmur was audible along the left sternal border. Left chest had decreased air entry and coarse crepitations. Chest roentgenogram revealed a small left hemithorax, shift of mediastimum towards the left, cavitation in the left upper lobe and fibrotic scaring in the left lobe. Right lung was oligemic with a small hilar shadow of right pulmonary artery. Two-dimensional echocardiography confirmed the diagnosis of TOF. Right pulmonary artery size was 10 mm. Left pulmonary artery could not be visualized. Angiocardiographic findings favoured the diagnosis of TOF, while the pulmonary angiogram showed absent left pulmonary artery.

Patient was operated upon and total correction of tetrology complex was done. Right pulmonary artery was of adequate size. There was no evidence of left pulmonary artery. Post operatively patient developed pulmonary edema and blood stained secretions were aspirated from the left bronchus. He was treated with diuretics and inotropes. After 72 hours of mechanical ventilation he was successfully weaned off and was discharged from the hospital on fifteenth postoperative day.

### DISCUSSION

Unilateral absence of pulmonary artery is an uncommon lesion. This condition was first reported by Fraentzel in 1868[6]. Pool reviewed world literature and collected 98 cases of UAPA. Embryological development of this condition has been explained by the atresia of the proximal sixth aortic arch, or by defective septation of the truncus arteriosus[2]. The affected lung is supplied from the bronchial arterial bed and anomalous systemic collaterals of intercostal and brochopulmonary origin[3].In the absence of bronchopulmonary anomalies of the affected side, patient may remain asymptomtic. Main clinical symptoms of isolated UAPA are recurrent respiratory infections, dyspnea on exertion and haemoptysis[4].

Correspondence:
Dr. Abid Aslam Awan,
Department of Cardiac Surgery,
National Institute of Cardiovascular Diseases, Karachi

Plain chest X-ray shows an asymmetric thorax with narrowed intercostal spaces on the affected side. Heart and the mediastimum are shifted to the affected side and there are unilaterally decreased vascular markings[5,8]. However, cardiac catheterization and selective central pulmonary angiography is mandatory for establishing the definitive diagnosis[1]. Aortography may reveal enlarged bronchial arteries and dilated internal mammary artery on the affected side [4].

In UAPA all the venous return goes to one lung with patent pulmonary artery. Initially the lung can accommodate the total cardiac output without much change in pulmonary vascular resistance, but with the passage of time pulmonary hypertension develops. If there is associated left to right shunt, pulmonary hypertension develops at an earlier stage. Disturbance of ventilation/perfusion (VA/Q) occurs in UAPA. VA/Q is decreased on the side where the pulmonary artery is patent and the lung receives the entire right ventricular output in the contralateral lung. Where the pulmonary artery is absent, the VA/Q is increased and therefore, the lung represents a dead space8. Only little oxygen uptake, 5% to 9% occurs in the lung, supplied from collateral systemic arteries[7]. A major disturbance of VA/Q may occur in UAPA patients during exercise and after surgery[8]. Sometime even life threatening pulmonary edema may develop.

Since all the gas exchange functions are maintained by one lung, its function can be easily compromised during or after surgery. In patients of TOF with UAPA, the risk of total surgical correction is high. After reconstruction of right ventricular outflow tract, all the cardiac output goes to the lung with patent pulmonary artery. Since preoperatively the lung is oligemic it cannot accommodate the increased blood volume leading to pulmonary edema. This was seen in our case. Previous shunt operation may be helpful in preparing pulmonary vascular bed for the post- correction load acceptance.

## REFERENCES


1. Pool PE, Vogel JHK and Blunt SG Jr. Congenital unilateral absence of a pulmonary artery. The importance of flow in pulmonary hypertension. Ann J Cardiol 1962: 110-706.
2. Kacera V. Fiser B. Tuma and Hucin B. Unilateral Absence of Pulmonary Artery: a report on 19 selected clinical case. J. Thorac cardiovasc Surg 1982:30:152-158.
3. Grum CM. Yaranl JR. Cook SA, Cordesco EM and Tomashefski JF. Unilateral hyperlucent lung. Non-invasive diagnosis of pulmonary artery agenecis. Angiology 1981;32:194.
4. Byrad RIR and Bloom DL: Absence of the right pulmonary artery as a cause of hemoptysis. J. Thorac Cardiovas Surg 1970;59:264.
5. Somora RF, Edwards JE. Anatomic identification of so-called absent pulmonary Artery. Circulation 1978;57:3.
6. Wyman SM. Congenital absence of a pulmonary artery: It demonstration by roentgenography, Radiology 1954;62:321.
7. Oakely-C, Glick-G, and Gredie RM: Congenital absence of a pulmonary artery. Am J Med 1962;34:264.
8. Elder JC, Brofman BC, Kohn PM and Chorms BL. Unilateral pulmonary artery absence or hypoplasia. Radiographic and cardiopulmonary studies in five patuebts. Circulation 1958;17:557.

# JUVENILE POLYPOSIS COLI

## A CASE REPORT

JAMSHED AKHTAR, ALI RAZA BROHI, ABDUL AZIZ.

*ABSTRACT:*
*Juvenile polyposis coli (JPC) is a rare polypoid lesion of gastrointestinal tract. A case of JPC in a twelve year old boy is reported in whom multiple polyps were present in distal half of colon. Total colectomy with ileo-anal anastomosis was performed. The interesting features related to the condition are also discussed.*

*KEY WORDS: Rectal bleeding, Juvenile polyposis coli, Colectomy.*

### INTRODUCTION

Polyps are common cause of rectal bleeding in children. Juvenile polyp is the leading variety. Usually they are single and are limited to rectum[1]. They are generally considered non-neoplastic lesions. Multiple juvenile polyposis (JP) is a rare condition[2]. The criteria for labeling a lesion as JP is not agreed upon. According to Sacahtello et al. ten or more colonic polyps or polyps throughout gastrointestinal tract or any number of polyps in the gastrointestinal tract with a family history are grouped under this heading[3]. Jass et al suggested that only five colonic polyps are enough to include it in this category[4] Giardiello et al are of the opinion that patients with as few as three polyps should undergo screening for colorectal neoplasia[5].

### CASE REPORT

A twelve years old male child weighing 30 kg was admitted with history of bleeding per rectum and rectal prolapse for the last two years. There was no family history of such problem. He was admitted to another hospital for 1 month where at proctoscopy multiple polyps were found and biopsy was taken that revealed juvenile polyp. The examination was unremarkable except for anemia. Perineal examination revealed patulous anus with partial rectal prolapse. The mucosa was studded with polyps of various sizes. Upper and lower GI barium series showed multiple polyps in distal colon. At colonoscopy polyps were found in rectum, sigmoid colon and descending colon. The polyps were of various size and few of them were pedunculated while others were sessile. Multiple polyps were removed for biopsy that revealed typical juvenile polyp. It was not possible to remove all the large sized polyps and it was also not certain that the patient would return for follow up so we decided to perform colectomy. After discussing with the relatives, patient was subjected to surgery. At laparotomy the lymph nodes of sigmoid mesentry were found enlarged. Multiple polyps were felt in distal colon. Small bowel was normal and so was the proximal colon. Total colectomy was done and ileoanal anastomosis was performed after performing mucosectomy of anal canal. Post operatively patient developed severe wound infection and dehiscence. After controlling infection, secondary suturing was performed. Repeat examination under anaesthesia revealed intact anastomosis that was easily calibrated.

In early post operative period patient developed frequent passage of loose stool up to 20 times a day with soiling and night time incontinence. Fluid and electrolyte imbalance was corrected. He also received multiple blood and plasma transfusions to maintain haemglobin level. Immodium was also started to decrease the motility of the gut. Gradually frequency decreased to 4-5 times per day and stool also became more solid. There was no soiling or night time incontinence. He was discharged after three months with 24 kg weight. The patient never returned for follow up.

Correspondence:
Dr. Jamshed Akhtar,
Asstt. Prof. of Paediatric Surgery,
National Institute of Child Health,
Karachi.

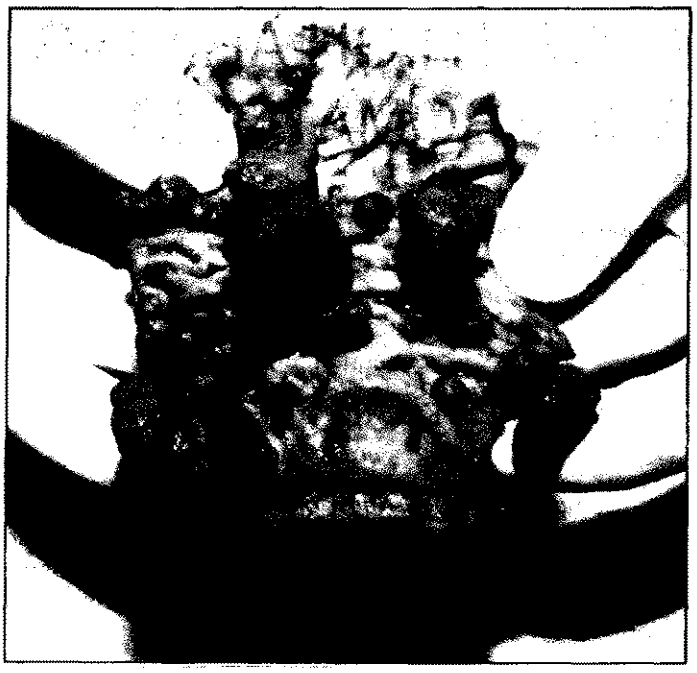

**Figure 1** Part of resected colon showing multiple polyps of varying sizes

The cut section of the colon revealed normal looking caecum, ascending colon and proximal transverse colon. Distal colon was studded with multiple polyps (Fig-I). The biopsy performed this time again revealed juvenile polyps. The lymph nodes showed inflammatory changes only.

## DISCUSSION

The neoplastic potential of JP is not known although increased incidence of colorectal carcinoma is reported in family members of both familial and non familial juvenile polyposis[6]. The probable sequence of events are transformation of JP to adenomatous polyp and ultimately development of adenocarcinoma. Kaschula was the first to report the co-existence of juvenile polyp and adenomatous polyp and presence of adenomatous epithelium in juvenile polyp[7].Third histologic variant is also described in which there is less stromal tissue and polyp shows villous pattern with lining of dysplastic epithelium[8]. There are also reports of varied morphology in solitary juvenile polyps[9].

The patients usually present with bleeding per rectum[10], rectal prolapse and anaemia. The management of these lesions is not only to keep the patients free of symptoms but also long term surveillance is required because of high risk of carcinoma developing at an early age[2]. Prophylactic colectomy has been suggested with rectal mucosectomy and ileo-anal pull through as primary procedure8. We also adopted the same procedure.

The post operative course was very difficult to manage in our patient. As expected patient developed loose motions and soiling a complication inherent to straight ileo-anal pull through procedure. The ileum gradually adapts and becomes dilated to accommodate the stool as happened in our patient. Various ileal reservoir procedures are described in an attempt to decrease the frequency of bowel movements and to lessen the chances of night time incontinence and soiling. These procedures are quite demanding and many complications are associated with them including difficulty in evacuation, pouchitis, small bowel obstruction, pelvic sepsis and loss of continence[11]. Another very important long term complication in these children in impairment of physical growth. Our patient lost 6 kg weight in three months. This aspect needs frequent follow up and dietary manipulations so that child could achieve normal physical parameters.

## REFERENCES


1. Jalihal A, Misra SP, Arvind AS, Kamath PS. Colonoscopic polypectomy in children. J Paediatr Surg. 1992: 27 (9), 1220-22.
2. Coburn MC et al. Malignant potential in intestinal juvenile polyposis syndromes. Ann Surg Oncol. 1995: 2, 935.
3. Sachatello CR, Hans IS, Carrington CB. Juvenile gastrointestinal polyposis in female infant: Report of a case and review of literature of a recently recognized syndrome. Surgery. 1974: 75, 107-114.
4. Jass JR, Williams CB, Bussey HJR et al. Juvenile polyposis--a precancerous condition. Histopathology. 1988: 13, 619-30.
5. Giardiello FM, Hamilton SR, Kern SE et al. Colorectal neo plasia in juvenile polyposis or juvenile polyps. Arch Dis Child. 1991: 66, 971-75.
6. Giliansky NH, Elliot MS, Price SK et al. The nutritional consequences and neoplastic potential of juvenile polyposis coli. Dis Colon Rectum. 1986: 29, 417-20.
7. Kaschula RO. Mixed juvenile, adenomatous and intermediate polyposis coli: Report of a case. Dis Colon Rectum. 1971: 14, 368-74.
8. Jarvinen H, Franssila KO. Familial juvenile polyposis coli: Increased risk of colorectal cancer. Gut. 1984: 25, 792-800.
9. Gryboski JD. All juvenile polyps are not benign. Am J Gastroenterol. 1986: 81, 397.
10. Vaiphai K, Thapa BR. Juvenile polyposis (coli)--High incidence of dysplastic epithelium. J Paediatr Surg. 1997. 32 (9), 1287-90.
11. Shilyansky J, Lelli JL, Drongowski RA, Coran AG. Efficacy of the straight endorectal pull-through in the management of familial adenomatous polyposis--A 16-years experience.

# NEUROFIBROSARCOMA OF THIGH ARISING IN A CASE OF NEUROFIBROMATOSIS

## A CASE REPORT

AYESHA RASHID*, ABID QAZI*, M. NAEEM KHAN**.

*ABSTRACT:*
*This is a report of 14 year old boy who had neurofibromatosis Type-I, with plexiform neurofibromas, multiple café-au-lait spots, papillomas throughout the body and a large diffuse lesion of left thigh. The latter had completely disabled him. Investigations revealed this lesion to be a case of neurofibrosarcoma, which is a malignant peripheral nerve sheath tumor. Details of management are discussed in this report. A brief review of literature is also presented.*

*KEY WORDS: Neurofibromatosis, palliative resection, malignant transformation.*

### INTRODUCTION

Malignant peripheral nerve sheath tumors (MPNSTs) encompass a wide unusual group of neoplasms with features of neural differentation[1]. Most neurofibrosarcomas are seen in the form of malignant transformations in cases of pre-existing neurofibromatosis Neurofibromatosis can be divided into two separate syndromes on the basis of genetics and clinical presentation, neurofibromatosis I (NF-I), called Von Recklinghausen's neurofibromatosis or peripheral neurofibromatosis and neurofibromatosis II (NF-II), also called bilateral vestibular schwannoma syndrome or central neurofibromatosis[2].

In NF Type-I there may frequently occur extensive cutaneous lesions, café-au-lait spots, axillary freckling plexiform neurofibromas, fibroma molluscum, hypo or hyperpigmented spots, xanthogranulomas and angiomas[3]. Neurofibrosarcomatous change is associated with NF Type-I. When the malignancy occurs it may be multifocal and has a high rate of recurrence. Patients with neurofibromatosis Type-I also have a high risk of other types of malignancy such as rhabdomyosarcoma, Wilm's tumor and non-lymphocytic leukemia[4]. Our patient highlights most of the features described above.

Correspondence:
Prof. M. Naeem Khan,
The Children Hospital, Islamabad.
Pakistan Institute of Medical Science, Islamabad.
*The Aga Khan University & Hospital,
**Department of Paediatric Surgery,

### CASE REPORT

A 14 year old boy was brought with complaint of having a mass in left thigh for several months which had suddenly increased in size, recently. As a result, the patient was unable to extend his knee or walk properly. On examination, the patient was lean built and had widespread café-au-lait spots on his body. There was marked axillary freckling, raised cutaneous lesions and areas of hypo-and-hyper pigmentation. The lesion on the thigh was hyper pigmented, bigger than a football, extending from the groin to the under surface of knee and calf. It was solid and appeared to be incorporated with the deep flexors

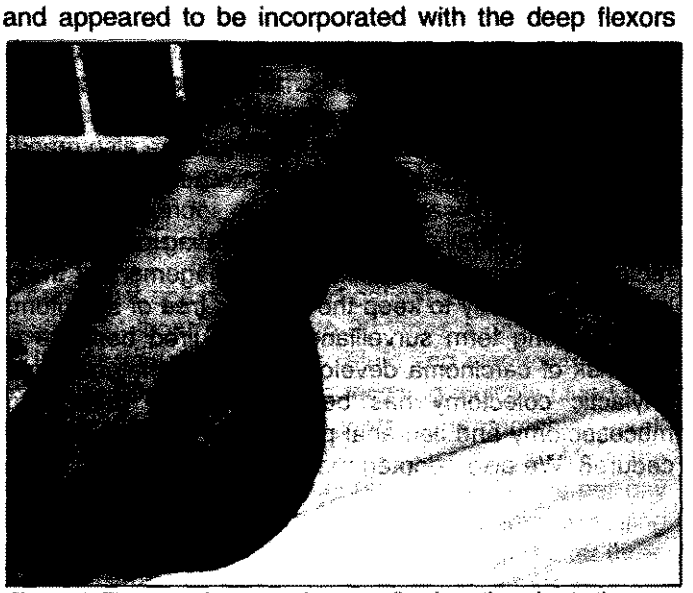

**Figure 1.** The tumor is seen to be extending from the grion to the upper calf

and adductors of the thigh (Fig-I). Since the boy was an orphan, there was no clear history of neurofibromatosis in the family. We carried certain investigations, including a magnetic resonance imaging (MRI) scan of the lesions that showed a massive soft tissue tumor with whorled appearance and was noted to be invading the fascial planes. There were several thick, enlarged cutaneous nerves but the blood vessels were spared (Figs-2a and 2b). There was no biopsy report of the lesion. Fine needle aspiration of the lesion was performed which revealed it to be a case of neurofibrosarcoma. Patient was further investigated with routine hematological tests and required pre-operative blood transfusions to correct anemia.

At operation the entire lesion was excised with considerable safe margins. Although it was insinuating in the inter muscular space and was plastered to the inter muscular fascia, it did not appear to invade the muscles. There were several worms like tortuous enlarged cutaneous nerves, which were removed en bloc with the lesion. There were few involved lymph nodes in the inguinal region which were also excised. The cut surface of the tumor revealed extensive central cystic degeneration of the tumor as if it had outgrown its blood supply (Fig-3). Histopathology of the tumor tissue confirmed this lesion to be a neurofibrosarcoma, which was presumed to be due to malignant change in a pre-existing neurofibroma. Post-operative progress was satisfactory and the patient was allowed to go home on the tenth post-operative day. In view of the extensive nature of growth and the pre-malignant potential of neurofibromatosis, the treatment was considered only to be palliative in nature and no further treatment was offered.

## DISCUSSION

Malignant peripheral nerve sheath tumors include a wide and unusual group of neoplasms with features of neural differentiation including neurofibrosarcoma. These tumors most commonly present as spindle cell neoplasms and it can be difficult to differentiate these from other spindle cell neoplasms, such as leiomyosarcoma, fibrosarcoma and synovial sarcoma[1]. MPNSTs are highly aggressive tumors and should be treated accordingly.

Various theories have been presented as far as the etiology of neurofibrosarcoma is concerned. The gene responsible for neurofibromatosis I has been located by using linkage analysis on the pericentromeric region of chromosome 17. The protein product of this gene, neurofibromin, is decreased in neurofibrosarcoma[4]. The neurofibromin gene seems to act as a tumor suppressor gene, and loss of both alleles is associated with formation of neurofibromas.

The question of malignancy after surgical intervention or of malignant transformation of a benign neurofibroma has been raised repeatedly in literature. Little evidence support the hypothesis that surgery induces sarcomatous transformation. Although most neurofibromas are benign, large tumors are known to harbour focal areas of sarcomatous change. Whether this represents a transformation or simply primary areas of sarcoma is unclear. However,

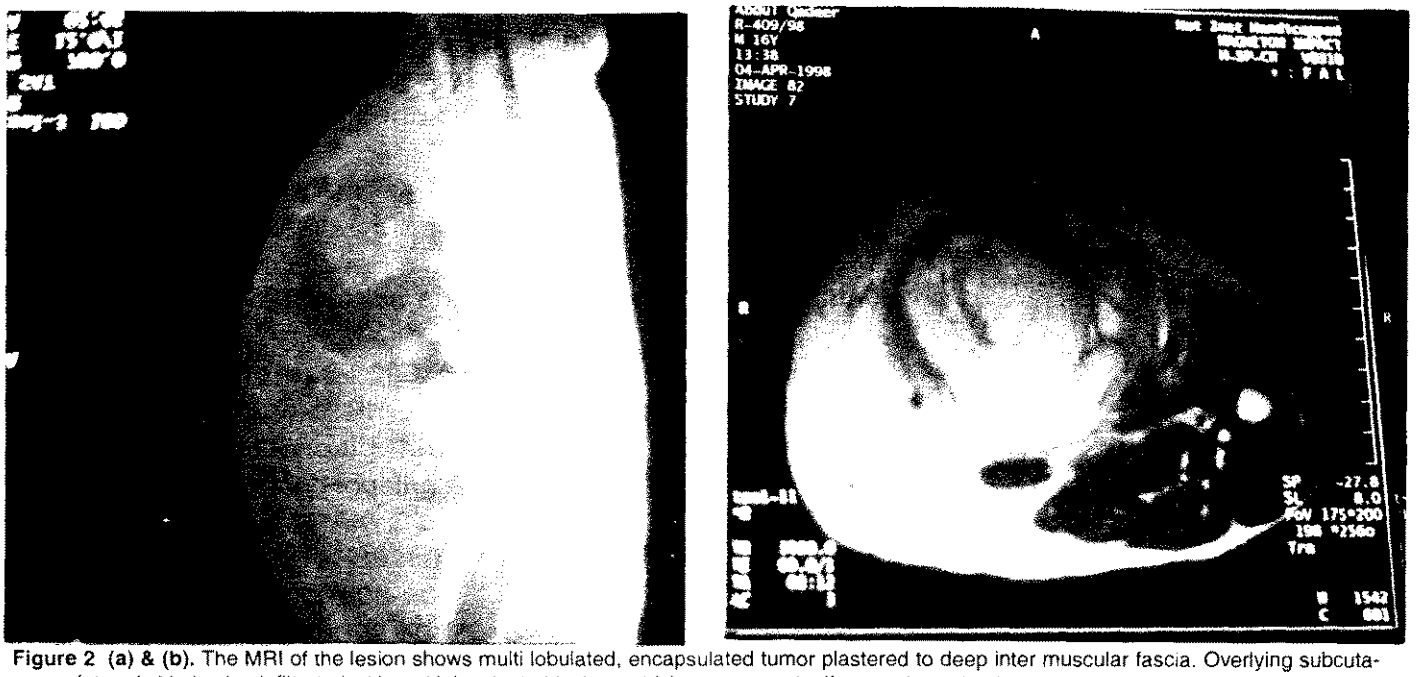


**Figure 2 (a) & (b).** The MRI of the lesion shows multi lobulated, encapsulated tumor plastered to deep inter muscular fascia. Overlying subcutaneous fat and skin is also infiltrated with multiple stippled lesions which represent plaxiform, enlarged cutaneous nerves

**Figure 3** The specimen shows complete removal of tumor with safe margins.

sarcomas associated with neurofibromatosis I tend to have a particularly malignant course. As emphasized earlier, neurofibroma is several times more common in patients with neuofibromatosis and develops at a much younger age. Thus, any increase in pain or rapid change in size of a neurofibroma or the rapid growth of a new tumor, as in this patient, demands prompt histologic diagnosis.

Strict criteria need to be applied in order to make the diagnosis of MPNSTs helpful features include contiguity with a nerve or an association with Von Recklinghausen disease. The use of immunohistochemical stains may also help to confirm the diagnosis. Markers such as S-100 protein, neuroifilament, epithelial membrane antigen and leu (CD 57) are frequently used to access neural differentiation in these neoplasms. In addition to the spindle cell pattern, MPNSTs may also display an epitheloid pattern[5].

MPNSTs are highly aggressive tumors and should be managed accordingly. Chemotherapy and radiotherapy are of limited value in this otherwise lethal sarcoma. If malignancy is suspected in a neurofibroma, the tumor should be radically excised en bloc, including the vascular supply, muscle and bone[2]. Individual tumors that are large or growing or cause pain or disfigurement should be removed. The treatment is by radical excision of the involved nerve and surrounding tissue. The tumors are often disseminated at diagnosis and have a high rate of local recurrence. The five year survival rate ranges from 40% to 45%.

## REFERENCES


1. Sangueza OP, Requena L. Neoplasms with neural differentiation a review Part II. Malignant neoplasms. An-J-Dermatopathol. 1998; 20(1) 89-102.
2. Rowe M, Paediatric Surgery Vol. Two, Fifth edition. 1998 pages 1915-1916.
3. Batool S, Siddique A, Patel A, Abbasi Z, Qazi A and Khan N. An experience with obvious external manifestations of neurofibromatosis & review of its etiology & other manifestations. Specialist, Pakistan J. Med Sci, 1995 II, (4) 335-340.
4. Riccardi V.M. Molecular biology of the neurofibromatoses, Sem Dermatol 1993, 12.266.
5. Neifeld JP, Prognostic features of pediatric soft tissue sarcomas, Surgery 1985, 98, 93.