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THANJAVUR

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CME PROGRAMME AT IMA NIDAMANGALAM - MANNARGUDI

IMA Nidamangalam - Mannargudi Branch in association with Meenakshi Hospital,Tanjore conducted a CME on 17-02-2024.The event was presided by Hony.Office Bearers: President - Dr.C.Manavalagan, Secretary - Dr.G.Saravanakumar, Treasurer – Dr.S.Vigneshwaran. Dr.R.Nirmala Sr.Consultant - Dept of Obstetrics and Gynaecology and Dr.N.Arunkumar Sr.Consultant - Dept of Neurosurgery, Meenakshi Hospital,Tanjore delivered the talk.

CME PROGRAMME AT IMA THIRUVARUR

IMA Thiruvarur Branch in association with Meenakshi Hospital,Tanjore conducted a CME on 18-02-2024. The event was presided by Hony.Office Bearers: President - Dr.E.M.G.S.Gopalakrishnapandian, Secretary - Dr.A.Ravindrababu, Treasurer - Dr.S.Anish Sunder Narayanan. Dr.M.Arun,Consultant - Dept of Respiratory Medicine and Dr.P.Isaac Richards, Consultant - Dept of ENT of Meenakshi Hospital,Tanjore delivered the talk.

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FROM THE EDITOR'S DESK

Dear Readers,

On this Year's International Women's Day, our editorial desk takes a moment to reflect on the pivotal role that women play in healthcare worldwide. From pioneering research to frontline caregiving, women have long been at the forefront of advancing medicine and improving patient outcomes.

Women's Day serves as an opportunity to celebrate the achievements of women in healthcare while also acknowledging the challenges they continue to face. Despite significant progress, gender disparities persist in various aspects of the medical field, including representation in leadership roles, access to resources, and recognition for their contributions.

As we honor the countless women who have made indelible marks on the healthcare landscape, we must also renew our commitment to addressing the barriers that hinder their full participation and advancement. It is imperative that we work towards creating inclusive environments that empower women to thrive professionally and personally.

Moreover, let us use this occasion to advocate for policies and initiatives that promote gender equity in healthcare, from supporting women's career development to ensuring equal pay and opportunities for leadership positions. By fostering a more diverse and inclusive healthcare workforce, we can better address the diverse needs of patients and communities around the world.

As We keep ourselves committed to advancing the field of healthcare, we stand in solidarity with women in healthcare and reaffirm our dedication to amplifying their voices, recognizing their contributions, and championing their rights.

Happy International Women's Day to all the remarkable women shaping the future of healthcare.

Learn – Share– Progress.,

Dr. SASIKUMAR SAMBASIVAM
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SPOTTER - 02

quiz

Quiz prepared by

Dr. Sasikumar Sambasivam, MBBS.,DNB., MNAMS.,PDCR.,
Sr.Consultant & HOD - Radiation Oncology,
Meenakshi Hospital

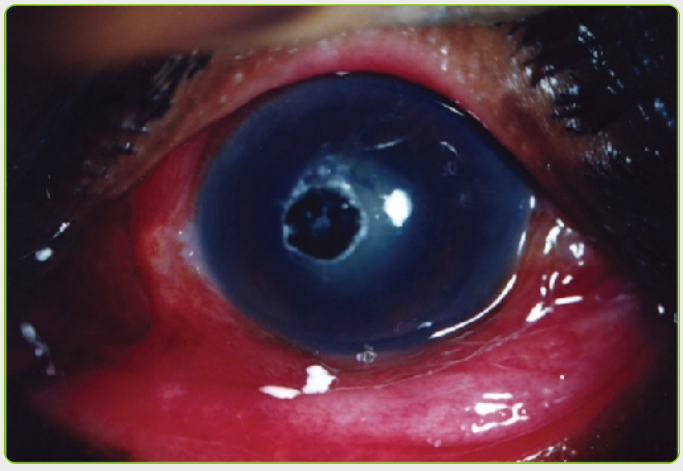
Quiz Details:

In FIGO (2018) Staging for Carcinoma Cervix: Presence of Para aortic Nodes is staged as:

A. IIIB
B. IIIC1
C. IV B
D. IIIC2

- Send your answer within **20th July 2024.**
- Write your full name, qualification and place of practice.
- Answer can be sent through whatsapp - **9599038089**
- For enquiries you can contact the above mentioned phone number.

SPOTTER - 01



Quiz Details:

Identify the Condition

A). Corneal Foreign Body
B). Fungal Corneal Ulcer
C). Post-traumatic Iris Prolapse
D). Viral Keratitis

Answer : B. Fungal Corneal Ulcer - It Is a Pigmented Fungal Corneal Ulcer Caused By Dematiaceous Fungi

last Issue quiz ANSWER

Congratulations...

WINNER OF THE LAST ISSUE QUIZ!

- **Tanjore** — K. Kirubananthan
- **Tanjore** — Dr. V.K.R. Palaniappan
- **Ariyalur** — Dr. S.Kandasamy
- **Tanjore** — Dr. Killivalavan
- **Samayapuram, Trichy** — Dr. T. Muthukumar
- **Saliyamangalam, Tanjore** — Dr.Suganya
- **Pudukkottai** — Dr. S. Ramraj

A RARE CASE REPORT OF INTERSITIAL LUNG DISEASE DUE TO SURFACTANT METABOLISM DYSFUNCTION, PULMONARY 2 (SMDP2)

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ABSTRACT: This is a case report of childhood interstitial lung disease due to uncommon mutation in SFTPC gene causing Pulmonary surfactant metabolism dystunction (SMDP2). A 3 month old male child presented to us with hypoxia and failure to thrive. Child was diagnosed to have rare mutation in SFTPC gene causing surfactant metabolism dysfunction. On treatment with hydroxychloroquine and steroid child is better now with adequate gain and normal work of breathing and saturation.

INTRODUCTION

Childhood Interstitial Lung Diseases are an uncommon group of heterogeneous disorders characterised by poor ventilation and impaired gas exchange with variable degrees of interstitial and alveolar inflammation and diffuse infiltrates on imaging. The early age of onset of clinical symptoms, progressive nature of disease and reporting of familial cases strongly suggests the presence of genetic factors involved in the development of this condition.

Pulmonary surfactant is a mixture consisting of phospholipids and 4 surfactant proteins – SP-A, SP-B, SP-C and SP-D, which are synthesized by alveolar type II pneumocytes. Specific inborn errors of surfactant metabolism have been identified resulting in Paediatric Interstitial Lung Diseases (ILD). The hydrophobic surfactant proteins SP-B and SP-C interact with the lipids to form a surface-active film at the air-liquid interface and reduce the surface tension, thereby preventing the alveoli and terminal bronchioles from collapsing.

Surfactant Protein B (SFTP-B) mutations are well described whereas the clinical presentation and outcome of lung disease associated with Surfactant Protein C (SFTP-C) is variable in view of its rare occurrence. Therapeutic options for SFTPC mutation disorders are also currently limited with lung transplantation being the ultimate treatment option.

CLINICAL DETAILS

A 3 months old male child, first born to non-consanguineous marriage presented with complaints of increased work of breathing and poor feeding of one month duration. Baby was delivered at our centre at late preterm gestation by emergency caesarean section in view of fetal distress with a birth weight of 2.3kg. Antenatal and Postnatal period was uneventful. Baby not to have poor weight gain with weight of only 2.9 kg. On admission, the baby had increased work of breathing with hypoxemia (SpO2 of 80-85% in room air). Baby appeared dull

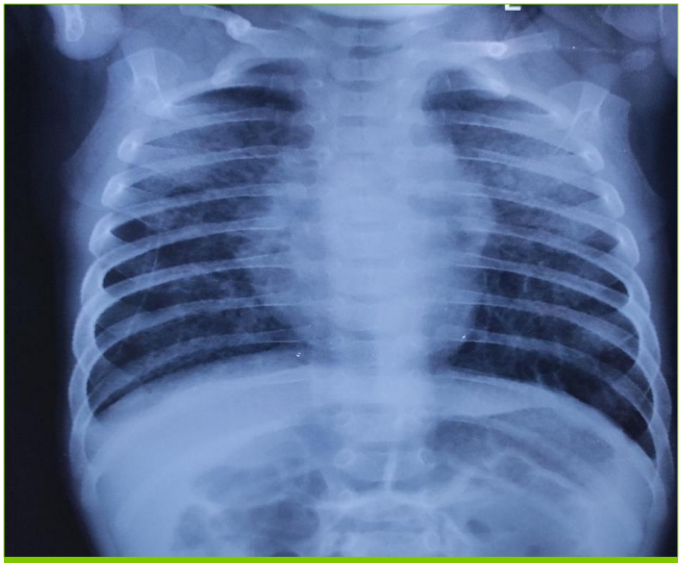

Fig 1 – Chest X-Ray at admission

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and dehydrated. On auscultation normal vesicular breath sounds were heard on both sides with no added sounds and Cardiovascular examination was normal.

The differential diagnosis considered were Congenital Cardiac disease, Respiratory disorders, Inborn errors of metabolism and feeding issues. Complete blood count and Blood gas was normal. Septic screen was Negative. Chest X-Ray (Fig 1) showed heterogenous opacification in bilateral upper lobes. Echo was normal. In view of persistent hypoxemia CT Thorax was done which showed bilateral symmetrical ground glass opacities, predominantly perihilar in location, with relative sparing of periphery of lungs, suggestive of Interstitial lung disease (Fig 2,3). Based on clinical examination and CT thorax

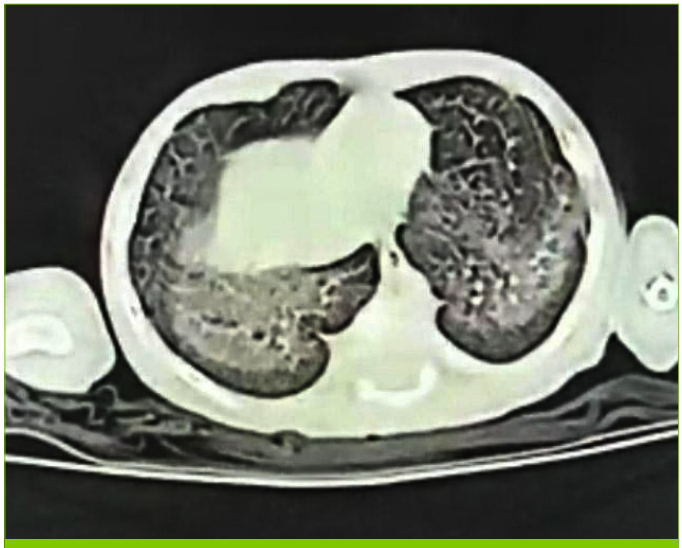

Fig 2 – CT Thorax at diagnosis

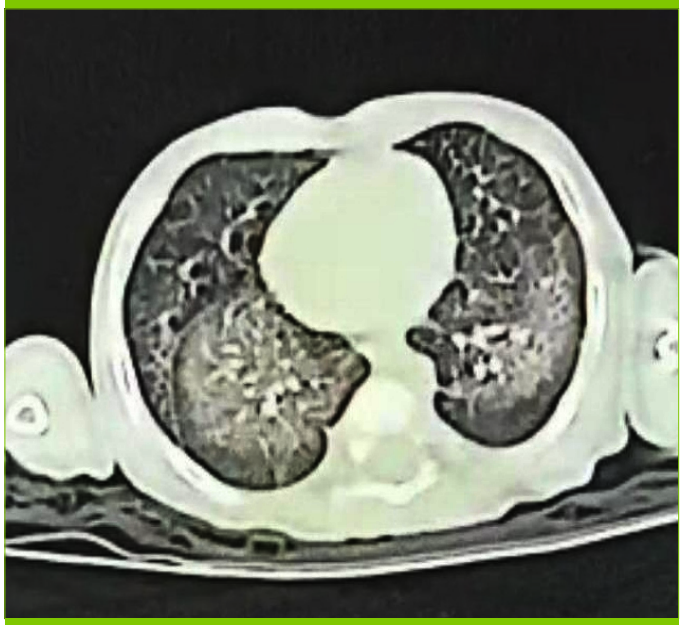

Fig 3 – CT Thorax at diagnosis

findings, congenital disorders of surfactant dysfunction was suspected and Whole genome sequencing was done, which showed heterozygous missense variant c.218T>C in Exon 3 of the SFTPC gene, suggestive of Pulmonary surfactant metabolism dysfunction type 2 (Fig 4). Child was admitted and initiated on oxygen support and orogastric feeding.

RESULT SUMMARY

PATHOGENIC VARIANTS CAUSATIVE OF THE REPORTED PHENOTYPE WERE IDENTIFIED

*** Correlation with clinical profile and family history is required**

Summary of Findings

Variants Potemtially Relevant to the indication for Testing :
The index patient is :
Heterozygous for a pathgenic variant in the SFTPC gene associated with SURFACTANT METABOLISM DYSFUNCTION, PULMONARY,2; SMDP2.

Carrier Status :
No pathogenic or likely pathogenic Variants were detected in the carrier gene list

Secondary Findings (AGMG gene List) :
No Pathogenic or likely Pathogenic (class 1/2) variants were detected in the ACMG gene list.

Fig 4 – Whole Genome Sequencing Report

Literature search done showed no definitive therapeutic options for SP-C deficiency. Child was started on treatment with monthly intravenous Methyl-Prednisolone at 10 mg/kg, oral Hydroxychloroquine 10mg/kg/day daily and oral Azithromycin 5mg/kg/day on alternate days. The decision to administer Pulse steroid therapy at a monthly interval was made to avoid systemic toxicity and to foster growth. Clinical improvement was commendable with respiratory distress and hypoxemia resolving over 72 hours. Feeding was established through an orogastric tube and weight gain was noted, following which baby was discharged. Oral feeding was gradually established and the child was administered routine immunisation. Child received monthly Intravenous methylprednisolone for 6 cycles along with Azithromycin and Hydroxychloroquine. Child gained weight gradually with normal work of breathing and oxygenation. Repeat CT thorax done after 6 months of treatment showed bilateral highly attenuating lung parenchyma with scattered simple cysts of varying sizes in bilateral lungs, remarkable improvement with normal lung parenchyma appearance (Fig 5,6). Child's respiratory parameters are within normal limits, with adequate weight gain and age-appropriate neuro-development. At present monthly steroids are stopped and the child is on daily Hydroxychloroquine.

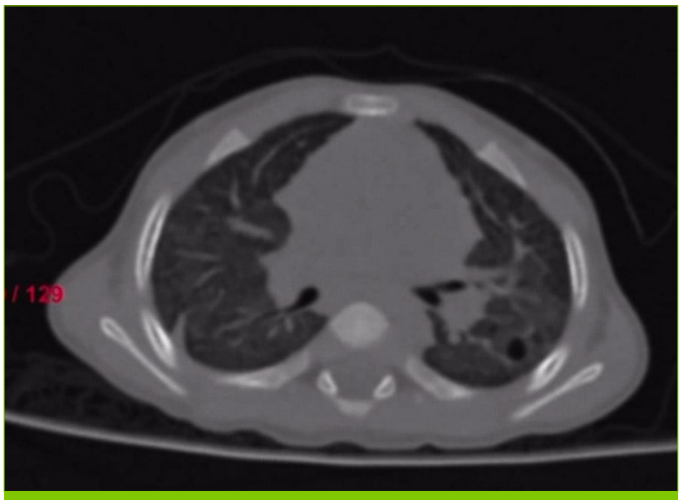

Fig 5 – CT Thorax after 6 months of treatment

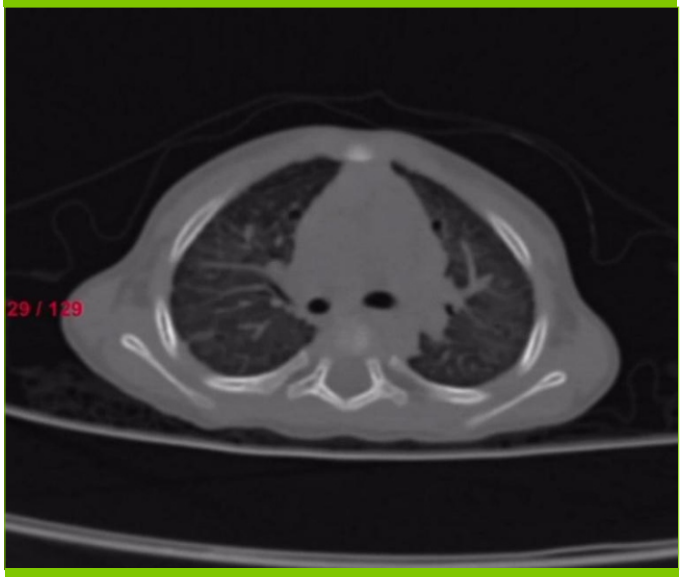

Fig 5 – CT Thorax after 6 months of treatment

DISCUSSION

According to our knowledge from the database, this is a rare case report of a severe respiratory failure caused by SFTP-C mutation.

Surfactant Protein C mutations resulting in Surfactant Protein C deficiency leading to respiratory failure during early infancy represent a rare entity. The condition was first described in 2001 with varying clinical outcomes. The onset of clinical presentation can start as early as the early neonatal period. Children who survive infancy usually suffer the sequelae of chronic lung disease[1].

SFTPC mutations result in the production of misfolded Protein SP-C, which accumulates within the alveolar cell type II, resulting in the activation of cellular stress responses and subsequent cellular injury and apoptosis[2].

The mutations identified so far are predominantly substitutions of highly conserved residues especially in the carboxy-terminal region of the protein. This domain predominantly has chaperone activity, sustaining the alpha helical structure of the transmembrane domain and in this process it prevents the self-aggregation of SP-C, before it combines with proteins and phospholipids to form active surfactant[3].

There is no specific treatment for SP-C deficiency based upon uncontrolled studies, case series, case reports and unsystematic observations. Steroids, Hydroxychloroquine and Azithromycin are commonly used in SP-C-related ILD. Azithromycin has anti-amyloid properties and Hydroxychloroquine affects intracellular Pro-SP-C processing, although there is currently no clinical data available to support this concept. Replacement of external surfactant provides only a transient improvement in gas exchange and is largely ineffective in SP-C deficiency[4]. Rosen and Waltz reported successful treatment of a 5-month-old boy with chronic lung disease due to SP-C deficiency with hydroxychloroquine. The mechanism of action may be a mixture of the drug's anti-inflammatory properties and a possible inhibition of the intracellular processing of the precursor of SP-C. Long term treatment with hydroxychloroquine may interfere with this accumulation of pro-surfactant proteins within alveolar cells[5]. More studies are needed to understand the mechanism of action of hydroxychloroquine in SP-C disease. An important question is the duration of treatment. Lung transplantation is an option for children with end-stage ILD, but its use is limited by donor availability and poor outcomes.

CONCLUSION

This case report highlights the importance of screening for SFTPC mutation in children with ILD of unknown etiology. There is a great need for multicentre randomized controlled trials in order to determine the best regimens for existing drugs that seem to improve the course of the disease, as well as additional basic and translational research to identify and test newer agents that improve long-term prognosis.

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AMNIOTIC MEMBRANE TRANSPLANTATION IN THE HUMAN EYE - REVIEW ARTICLE

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INTRODUCTION

The ocular surface is an extremely sensitive and dynamic structure, the health of which is crucial for the optimal functioning of the eye. Any mechanical or chemical insult to it either from exogenous sources, i.e., chemical injuries by substances like acids and alkalis, or from endogenous factors, i.e., change in the amount and composition of the tear film due to severe dry eye states associated with conditions like Stevens Johnson syndrome (SJS), rheumatoid arthritis and other collagen vascular diseases ,can result in anatomical, physiological and optical dysfunction of the eye as a whole.

Transplantation of preserved human amniotic membrane (AM) can be considered one of the major new developments in surgery of the ocular surface. Although the first ophthalmological use of AM documented in the international literature took place almost 70 years ago, amniotic membrane transplantation (AMT) has only been performed in larger numbers of patients since 1995, with promising results.

PROCUREMENT

The AM is the innermost layer of the placenta, located next to the fetus. Histologically, it is a multilayer membrane approximately 0.02 to 0.5 mm thick. After serologically detectable pathogens (HIV-1/2; hepatitis B, C; HTLV-I/II; syphilis) have been ruled out, it is procured under sterile conditions during a Cesarean section, packed in a sterile environment, and swiftly prepared in a sterile working area. If professionally procured and preserved, AM's biological properties are retained, and as a result this natural matrix can be used to replace damaged stromal tissue of the surface of the eye.

MECHANISM OF ACTION

Several mechanisms of action are attributed to the AM's ability to help in healing and reconstruction of the ocular surface.

1.Mechanical

The AM acts as a biological bandage and shields the regenerating epithelium from the frictional forces generated by the blinking movements of the eyelids.

2.Promotion of epithelialization

The basement membrane of the AM closely resembles that of the conjunctiva and cornea especially with regards to its collagen composition. It thus serves as a substrate on which epithelial cells can grow easily.

3.Anti-fibrotic and anti-inflammatory properties

Fetal hyaluronic acid is an important constituent of the stromal matrix of the AM. This helps to suppress TGF . which inhibits proliferation of corneal, limbal and conjunctival fibroblasts. Differentiation of fibroblasts into myofibroblasts is also inhibited, thus reducing scarring. Anti-inflammatory effect of AM is driven by inhibition of expression of pro inflammatory cytokines from the damaged ocular surface, e.g., interleukin (IL) 1a, IL-2, IL-8, interferon-., tumor necrosis factor-., basic fibroblast growth factor and platelet derived growth factor.

4.Anti-angiogenic properties

5.Anti- microbial properties

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INDICATIONS FOR AMT

1. Reconstruction of the corneal surface

- Persistent epithelial defect with corneal ulceration
- Reconstruction of the surface of the conjunctiva
- Acute chemical burns
- Removal of epithelial or subepithelial lesions (band keratopathy, scars, tumors)
- Painful bullous keratopathy
- Partial or complete limbal stem cell deficiency (with stem cell grafting)

2. Reconstruction of the conjunctival surface

- Acute chemical burns and acute Stevens-Johnson syndrome
- Covering defects after removal of large conjunctival lesions (tumors, conjunctival intraepithelial neoplasia, scars, conjunctival folds parallel to the edges of the eyelids)
- Symblepharon, fornix reconstruction
- Anophthalmia
- Bleb revisions
- Scleral thinning
- Pterygium

CONCLUSION

AMT is used in acute ophthalmological care, to treat chronic diseases of the surface of the eye, and as a latest development, using tissue engineering, as a biomatrix to treat severe stem cell deficiency of the ocular surface. It provides practicing ophthalmologists with a particularly multifaceted instrument to tackle the challenges posed by disorders of the surface of the eye successfully. Controlled, randomized, multicenter, long-term trials involving large numbers of patients are needed in order to substantiate the clinically relevant potential of AMT in reconstructive surgery of the surface of the eye, which has been documented in many case studies.

The AM is proving to be a very versatile tool in the hands of the ophthalmologist, and the indications for its use are rapidly expanding as there is a better understanding of its properties. However, judicious use and appropriate patient selection is important for achieving optimal outcomes.

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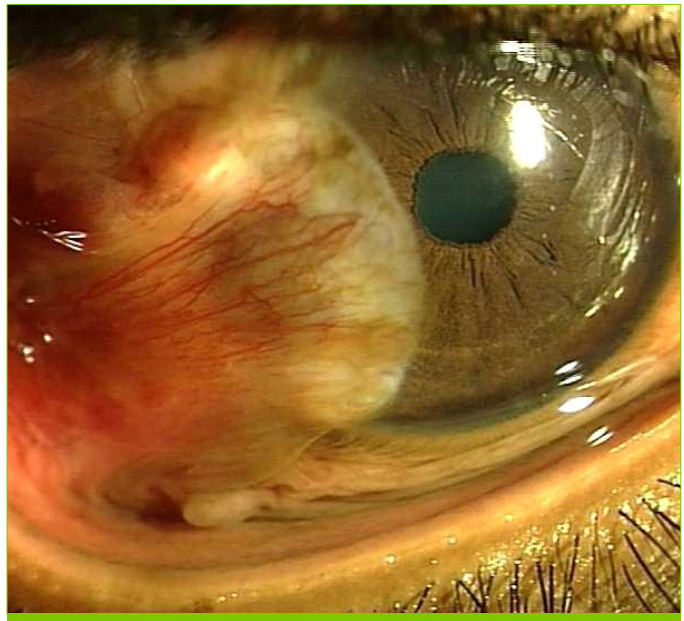

Fig 1 (a) PRE OP PHOTO OF PTERYGIUM

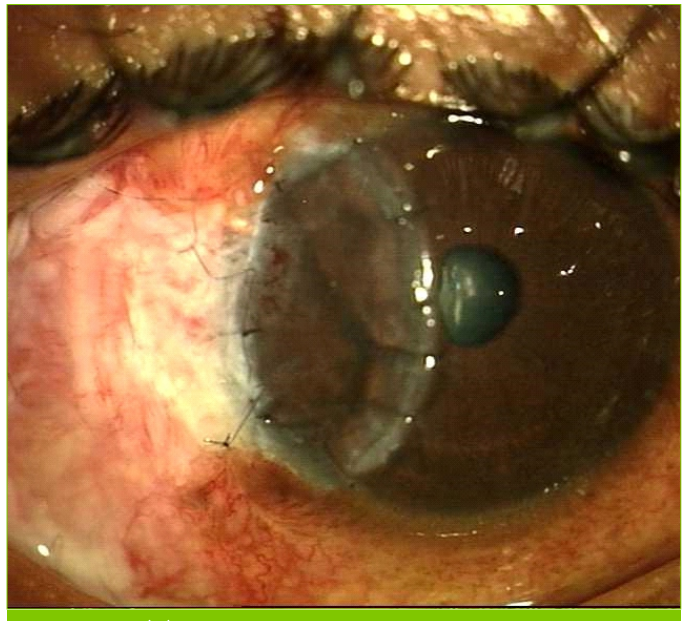

Fig 1 (b) POST OP PHOTO AFTER PTERYGIUM EXCISION AND AMG IN SITU

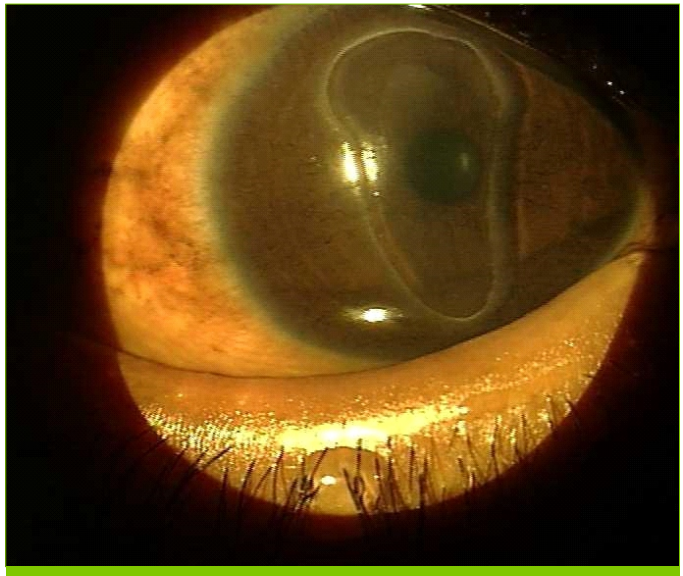

Fig 2 (a) PRE OP PHOTO OF SHIELD ULCER

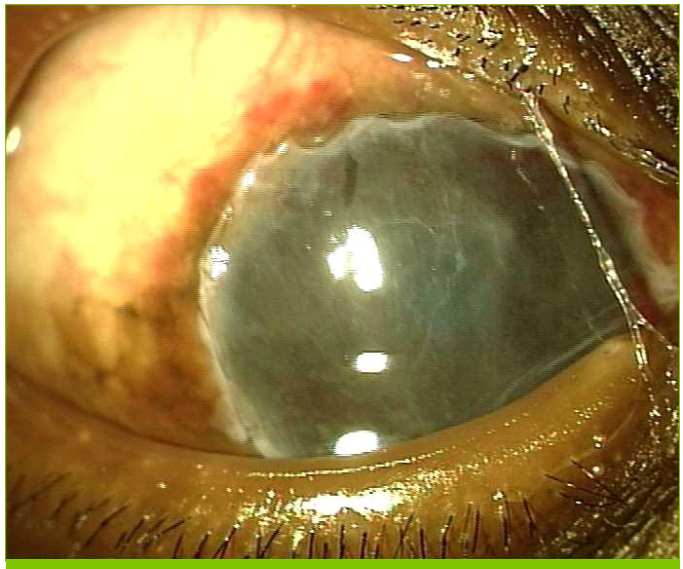

Fig 2 (b) AMG IN PLACE

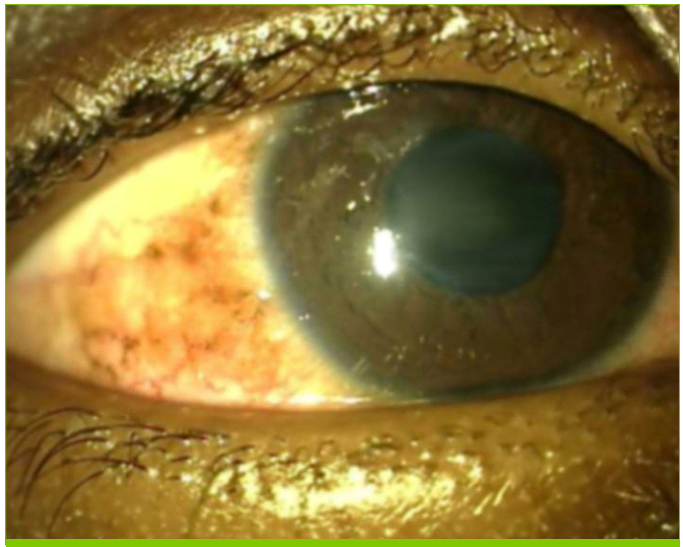

Fig 2 © HEALED ULCER

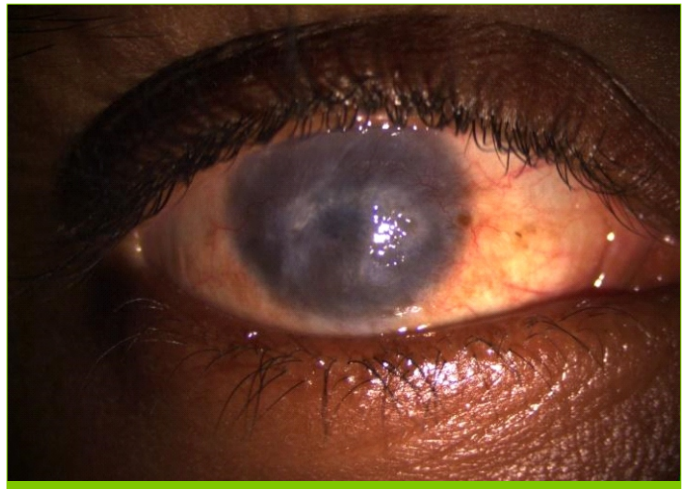

Fig 3 (a) PRE OP PHOTO OF CHEMICAL BURNS INDUCED CORNEAL OPACITY

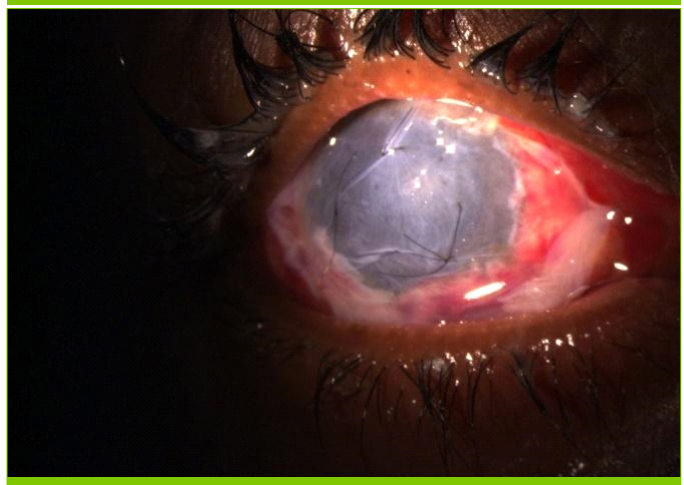

Fig 3 (b) AMG IN SITU

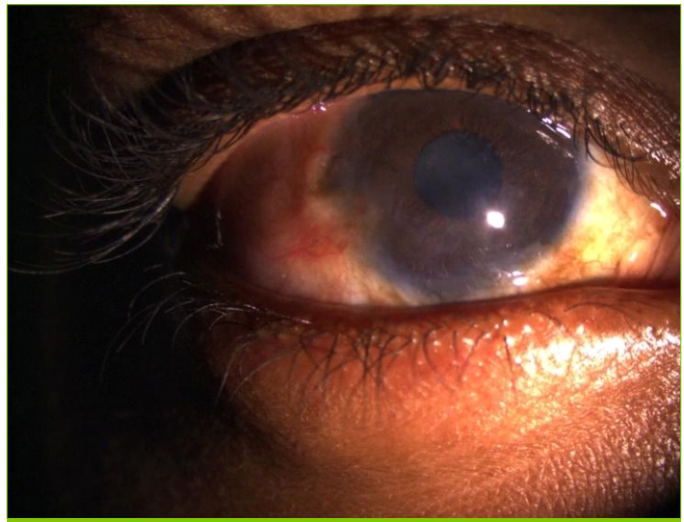

Fig 3 (c) POST OP - 2 Months of The Same Eye

MANAGEMENT OF COMPLEX PELVIC INJURIES -CASE SERIES

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ABSTRACT: Complex pelvis injuries following high energy trauma results in significant morbidity and mortality. Reported mortality from severe pelvic fractures ranges from 10% to as high as 50%. We have operated on nearly 50 patients with complex pelvis injury and functional outcomes were evaluated.

INTRODUCTION

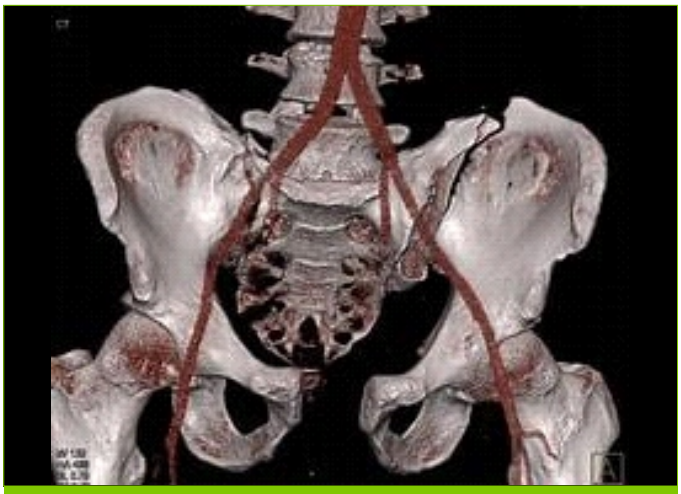

Fig 1: Complex Pelvis Fracture.

Fractures of the adult pelvis generally are either stable fractures resulting from low energy trauma and unstable fractures caused by high energy trauma. High energy pelvic fractures result most commonly from motor vehicle accidents, falls, automobile pedestrian encounters and industrial crush injuries. The potential complications of high energy pelvic fracture include injuries to major vessels, nerves of pelvis, major viscera such as intestines, bladder and urethra. Early mortality most commonly results from hemorrhage or closed head injury, late mortality occurs from sepsis or multiple system organ failure.

IMMEDIATE MANAGEMENT

The acute management of a patient with a pelvic fracture and unrelenting hemorrhage remains a challenge. A multidisciplinary approach with orthopedic surgeon, general surgeon and anesthesiologist is critical to optimizing outcomes. The initial trauma workup includes CT of chest and abdomen, supraumbilical peritoneal lavage, abdominal ultrasound. On recognition of unstable pelvic ring injury circumferential pelvic binder should be applied.

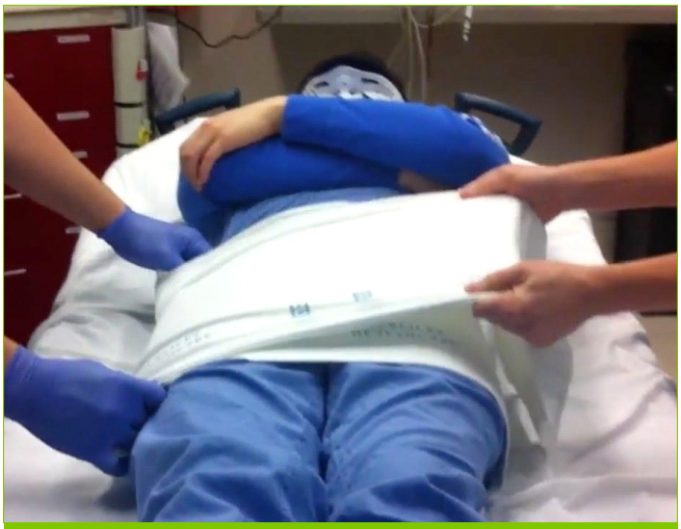

Fig 2: Emergency Pelvis Sheet Binding

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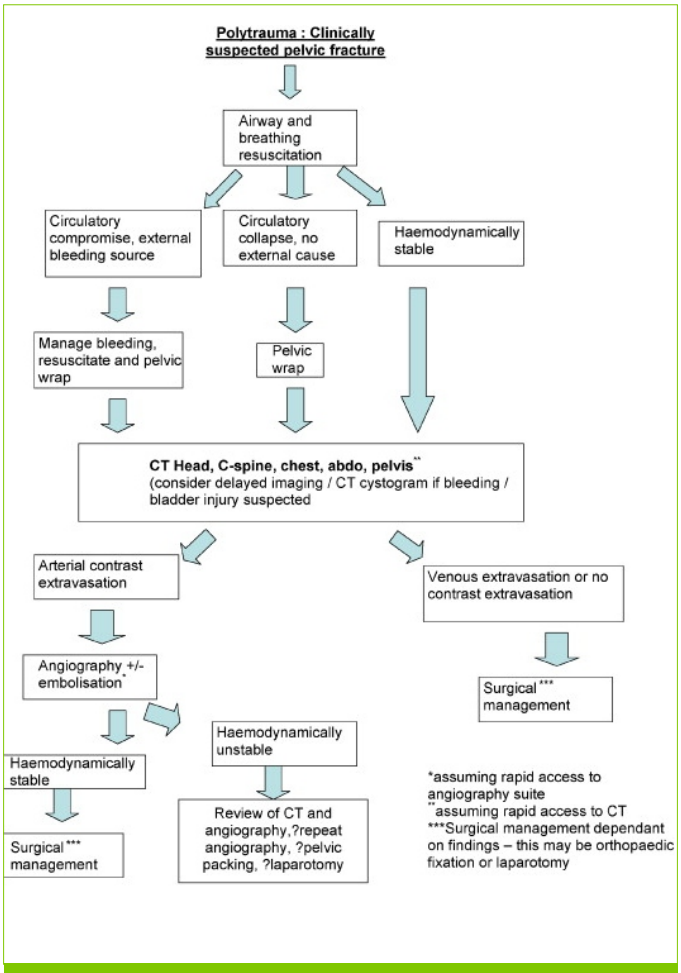
PERSISTENT HYPOTENSION

The patient with a pelvic ring injury, persistent hypotension after circumferential pelvic binding should be considered for arteriography. Hemorrhage frequently results from fracture surfaces and small vessels in the retroperitoneum. Only 5 to 10% patients with pelvic fractures bleed from arterial sources identified by angiography and are treated with embolization.

Open pelvic fractures are extremely difficult injuries to manage with a reported mortality rate up to 50%. Sepsis caused by fecal contamination is a major cause of mortality with this injury and immediate diverting colostomy is indicated in patients with perineal wounds.



Flow Chart of Complex Pelvis Injury Management

ANATOMY

The pelvis is composed anteriorly of the ring of pubis and ischial rami connected with the symphysis pubis. Posteriorly the sacrum and two innominate bones are joined. The primary restraints to external rotation of the hemipelvis are the ligaments of the symphysis, sacrospinous ligament and anterior sacroiliac ligament. Rotation in the sagittal plane is resisted by sacrotuberous ligament.

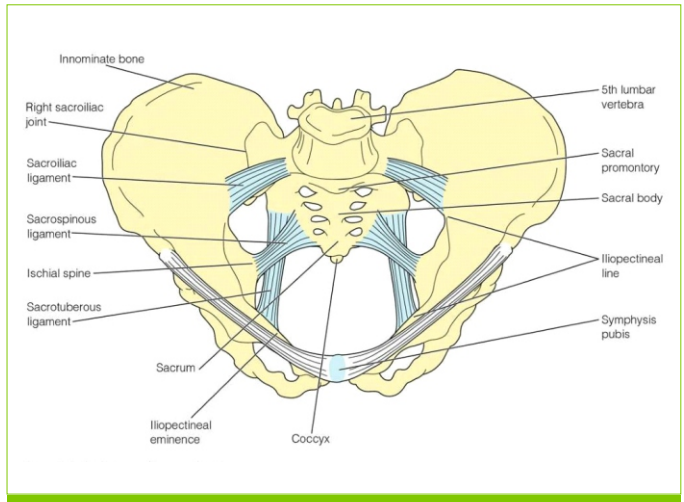


Fig 3: Pelvis Bone and Ligaments.

CLASSIFICATION

A – Stable [posterior arch intact]

B – Partially stable [incomplete disruption of posterior arch]

C – Unstable [complete disruption of posterior arch]

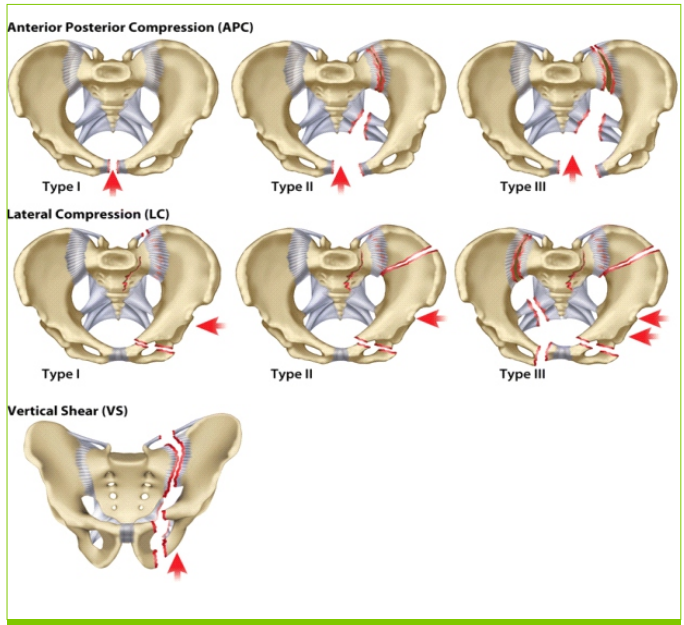


Fig 4: Classification on Force of Injury

RADIOGRAPHIC EVALUATION

The standard radiographic projections required for evaluation of pelvic fractures are anteroposterior view, 40* caudal inlet and 40* cephalad outlet view.

Inlet view shows rotational deformity or anteroposterior displacement of hemipelvis.

Outlet view shows vertical displacement of hemipelvis, sacral fractures and widening or fracture of anterior pelvis.

Widening of symphysis of more than 2.5 cm has been correlated with rupture of sacrospinous ligament and a rotationally unstable pelvis.

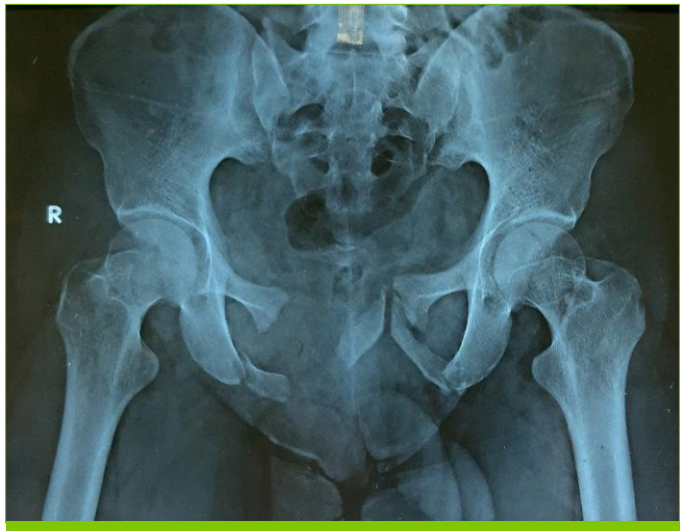


Fig 5: X ray of Complex Pelvis Fracture

PELVIC DAMAGE CONTROL

Closed reduction of the pelvis at admission.

External fixation

-wrapping pelvis with sheets with inner rotation and slight flexion of knees

-external fixator

-pelvic C clamp

-pneumatic antishock garment

Control of hemorrhage

-pelvic packing

-angiography

Control of contamination

-repair of genitourinary and rectal injuries

-debridement of necrotic tissue in the case of open injuries.

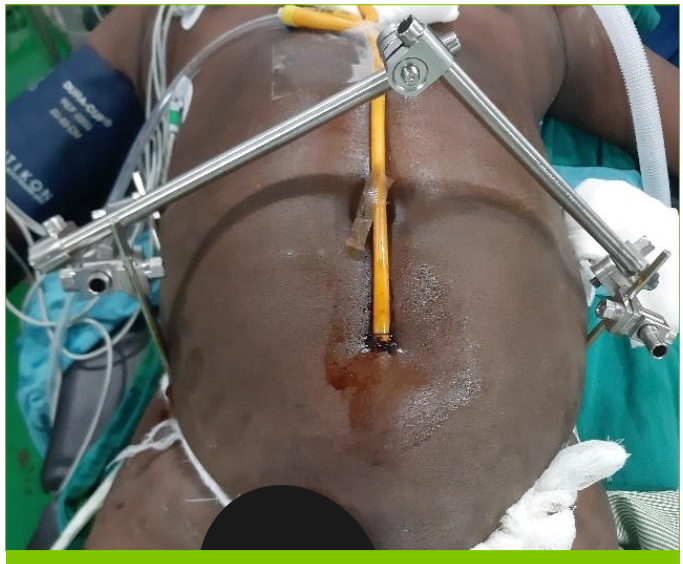


Fig 6: Emergency Pelvis External Fixation and Suprapubic Cystostomy.

MANAGEMENT

Stable nondisplaced pelvic fractures can be adequately managed with early mobilization and analgesics.

The significant mortality associated with nonoperative treatment of displaced, unstable pelvic fractures has led to a more aggressive operative approach.

Operative reduction and stabilization have been advocated for rotationally unstable fractures. Anterior plating of pubis symphysis disruption.

In vertically unstable pelvic injuries posterior fixation requires to regain vertical stability.

In rotationally and vertically unstable pelvic fractures, both anterior and posterior pelvis fixation is mandatory.

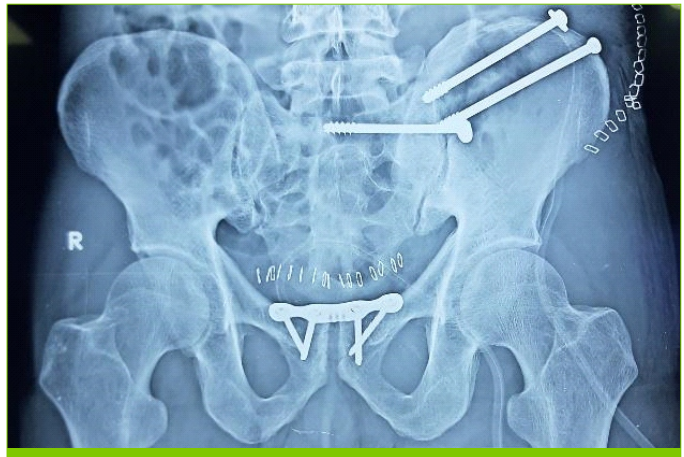


Fig 7: Fixation of SI joint, Iliac wing and Pubis Symphysis.

Neurological injury occurs with 30% of transforaminal sacral fractures.

For sacral fractures, sacroiliac joint disruptions posterior screw fixation into sacral body, trans iliac rod fixation, tension band plating of posterior iliac crest, anterior plating of sacroiliac joint.

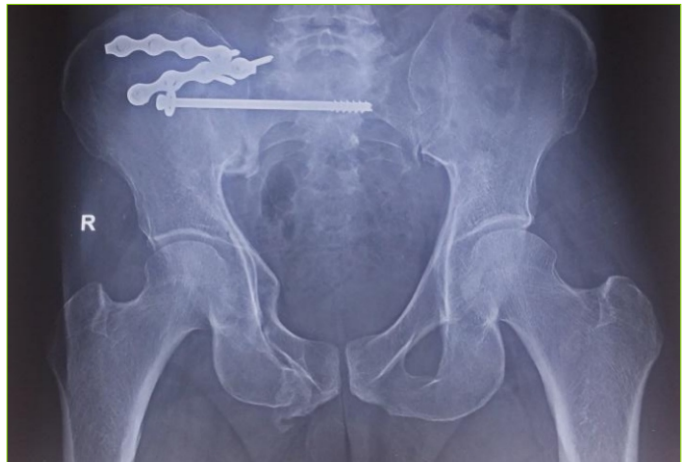


Fig 8: Fixation of SI joint with both Plating and Compression Screwing.

DISCUSSION

We have managed nearly 50 patients of complex pelvis injury with associated other organ injury. In which associated head injuries in 15 patients managed nonsurgically in 5 patients and surgical treatment in 10 patients. Associated abdominal organ injuries in 12 patients managed nonsurgically. In patients with open book pelvis injuries associated urethral injuries in 8 patients managed surgically.

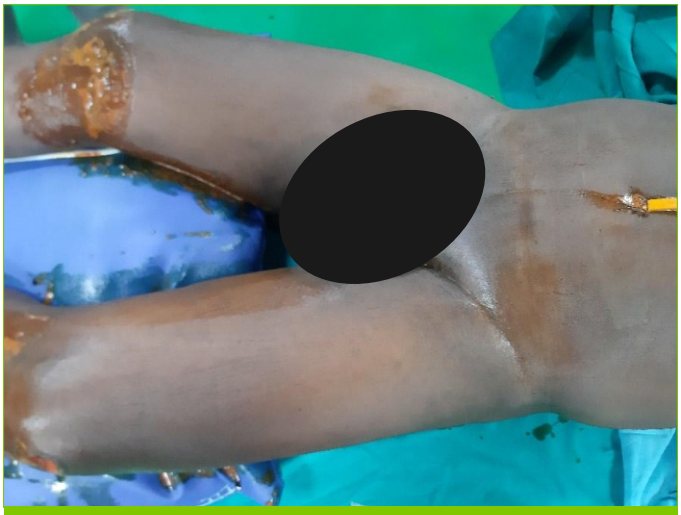


Fig 9: Pelvis associated with Urethral Injury.

Emergency resuscitation management of complex pelvis injuries with ABC protocol were followed. In 12 patients with unstable hemodynamic were intubated and resuscitated subsequently.

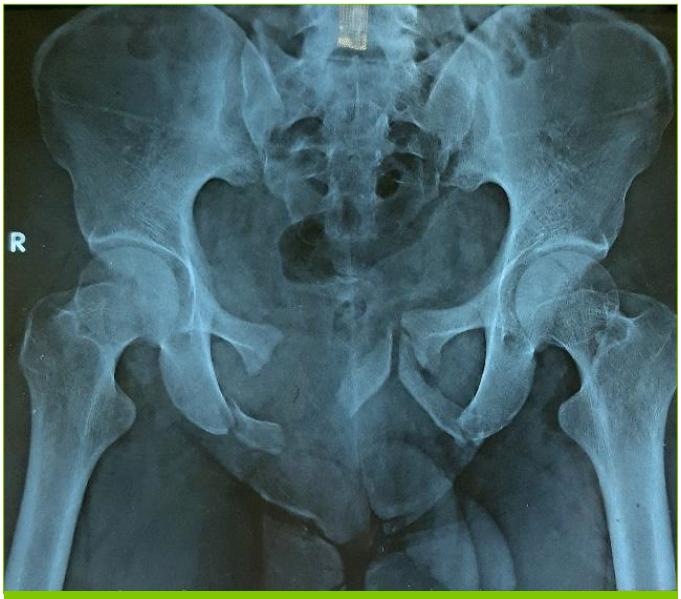


Fig 10 : Bilateral both Anterior and Posterior Pelvis Injury.

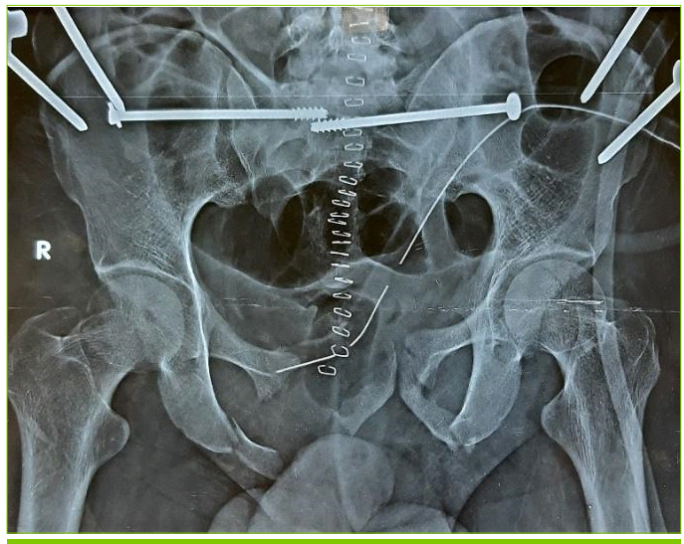


Fig 11: Stage 1 – Posterior Screw Fixation and Pelvis External Fixation.

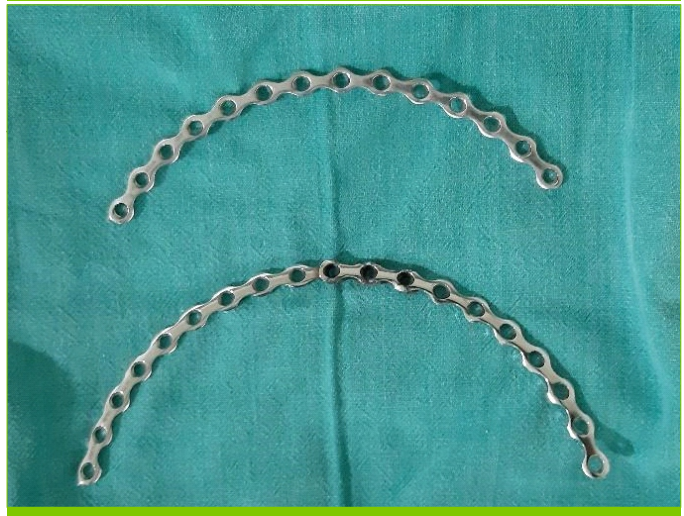


Fig 12: Custom made United Dual Plate.

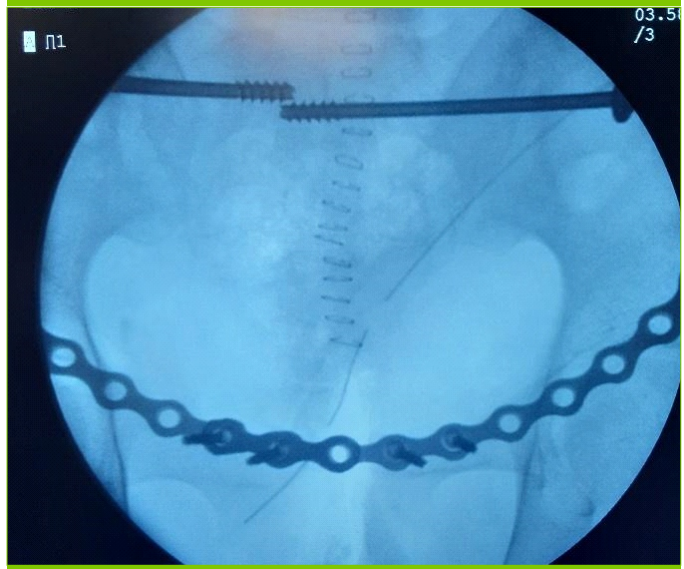


Fig 13: Stage 2 - Bilateral Anterior Pelvic Fixation with Custom made Dual Plate.

Mobilization was started on the 8th week with walker support. Stressful activities were allowed by 12th week.

Functional outcome was evaluated for pain relief, stair climbing, sitting cross leg and work regaining activities.

Complications which we have encountered are sciatic nerve injury [2], vascular injury [1], inadequate fracture reductions [3], delayed fracture union [5] and implant loosening [2] patients.

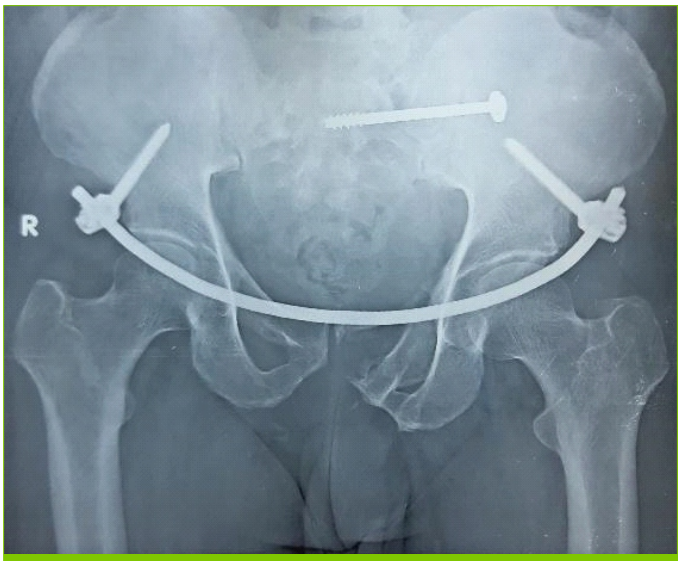


Fig 14: Minimal Invasive Percutaneous Fixation.

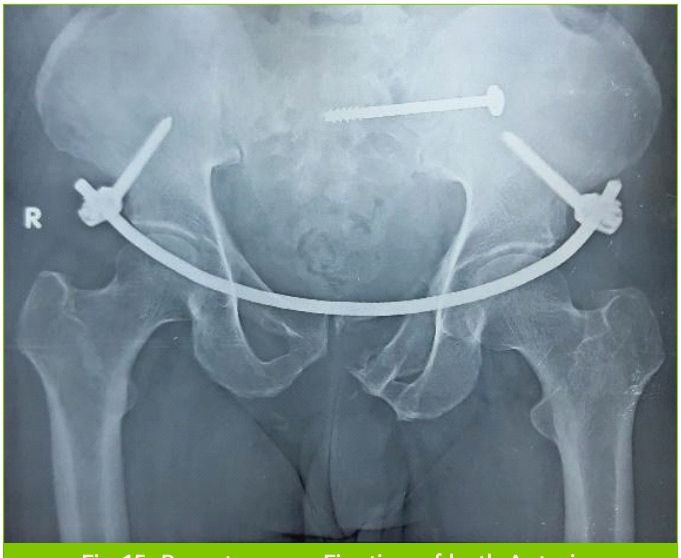


Fig 15: Percutaneous Fixation of both Anterior and Posterior Pelvis.

Fig 16: Functional Outcome of Pelvis Fixation.

Fig 17: Functional outcome of Complex Pelvis Fixation.

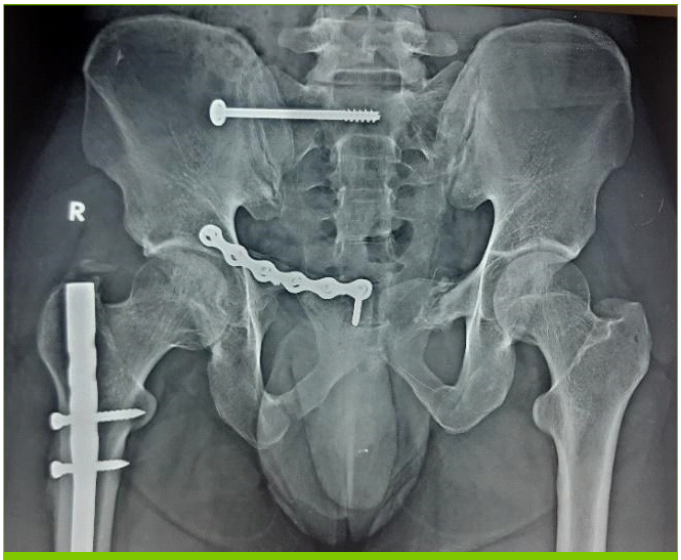


Fig 18: Complex Pelvis and Femur Fixation.

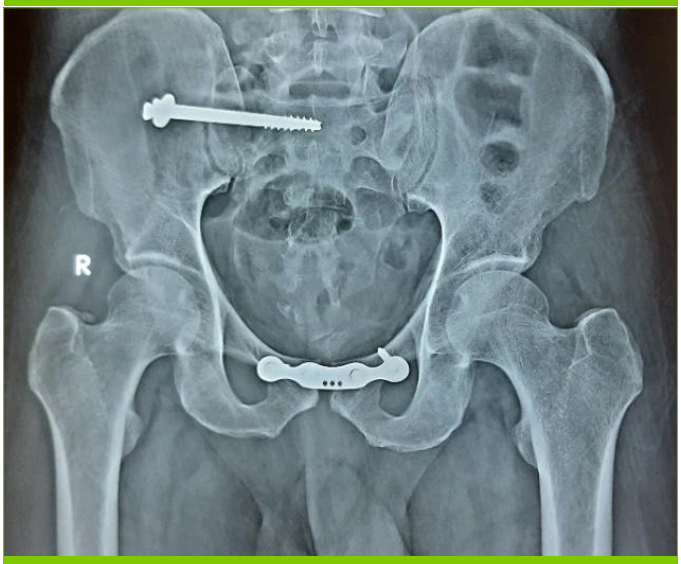


Fig 19: Both Anterior and Posterior Pelvis Fixation.

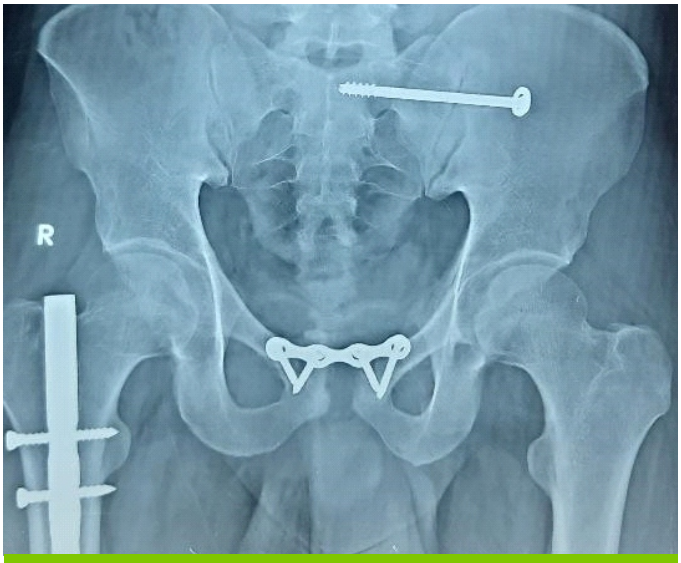


Fig 20: Complex Pelvis and Femur Fixation.

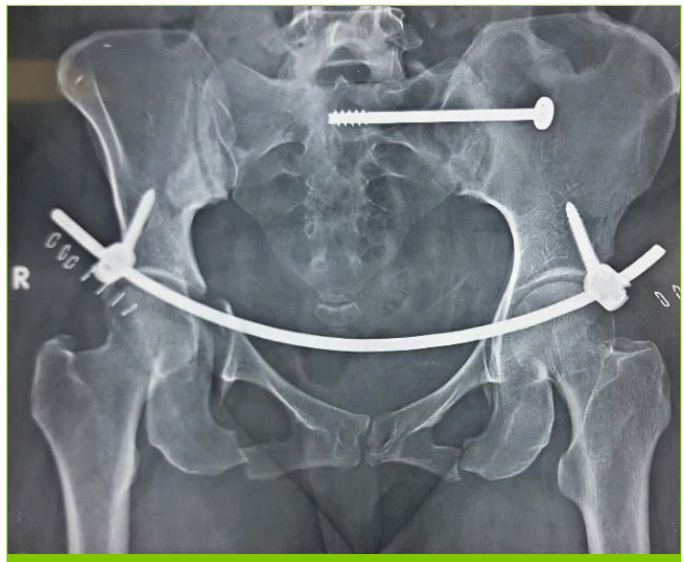


Fig 21: Percutaneous Pelvic Fixation.

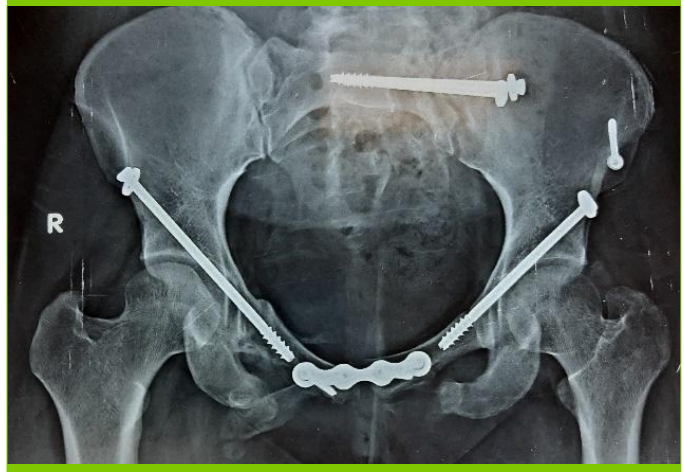


Fig 22: Complex Pelvis and Acetabulum Fixation.

Fig 23: Functional outcome of Pelvis Fixation.

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SCOLIOSIS CORRECTION SURGERY IN RETT SYNDROME

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INTRODUCTION

Rett syndrome is a progressive neurodevelopmental disorder predominantly affecting females and is associated with a high incidence of scoliosis and epilepsy. In this case report, I describe a 14-year-old girl with Rett syndrome with neuromuscular scoliosis and its correction surgery.

CASE REPORT

Miss ABN is a 14-year-old girl who presented to me in the clinic with complaints of deformity in the back and inability to sit and stand straight. The deformity was initially noted at 10 years and has gradually worsened since then. The parents noticed listing to the right side while sitting and standing. She had a normal antenatal history and postnatal history. She was born full term and had achieved milestones according to her age till 1 to 2 years of age. After that, there was a regression of her milestones. At 14 years old, she was able to sit and walk in the house unsteadily with a spinal list. Her mental development is grossly impaired with poor speech and motor skills. She has a history of epilepsy and was on anti-epileptic medications. She was grossly underweight and was measuring only 25 kg during the first consultation. There were no other organ dysfunction.

Clinical examination demonstrated a rib and loin hump on the left side. There were no neurocutaneous markers. No gross neuro deficits could be demonstrated as a detailed neurological examination was not possible. X-ray of the whole spine sitting demonstrated a long C-shaped levoscoliotic curve with a Cobb's angle of 80 degrees. MRI and CT did not demonstrate any bony or spinal cord abnormalities. Traction X Ray demonstrated partial correction of the curve suggesting some curve flexibility. There was no limb length discrepancy or pelvic obliquity on sitting. The patient was counseled for surgery as the curve was more than 50 degrees. Since she was grossly underweight, she was advised nutritional supplements to gain weight. However, she gained only 3 kg of weight to 28 kg even after following her for 3 months.

She was worked up for scoliosis correction surgery. The surgery was performed under TIVA(Total Intravenous Anesthesia) to aid neuromonitoring. Intraoperative neuromonitoring technology was utilized to monitor the spinal cord function throughout the procedure. MEPs, SSEPs and EMGs were continuously monitored throughout the procedure. Exposure was performed from T3 to S1. Extreme care was taken to minimize the blood loss. Through subperiosteal dissection, all bony landmarks were exposed. Pedicle screws were placed on both convex (L5, L4, L2, T11, T9, T7, T4, T3) and concave sides (L5, L4, L3, L1, T12, T10, T8, T6, T4, T3) using freehand technique. After the insertion of screws on both the concave and convex sides was complete, a slightly over-contoured rod was inserted on the concave side. Rod reduction was facilitated using rod reducers sequentially. Rod derotation was performed to regain the sagittal profile. Then, a convex rod was placed and reduced sequentially with reducers. Final compression and distraction were done to achieve the final balance Neuromonitoring was performed continuously throughout the procedure and no drop in signals was observed till the end of the procedure. Bone

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decortication and fusion bed with local autograft were created over the instrumented region. Intraoperative blood loss was around 300 ml and no intraoperative blood transfusion was needed. No major postoperative complications were encountered. The patient was made to mobilize from postoperative day 3. She was discharged home on POD 8. At 3 months follow-up, she is doing extremely well with good sagittal and coronal balance while sitting and walking. Postoperative X-ray demonstrated good correction of the deformity.

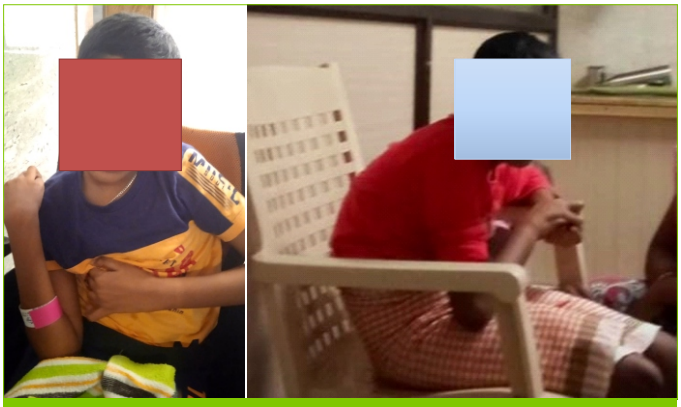

Figure 1: Preoperative clinical photography showing slouching attitude and Right-sided list.

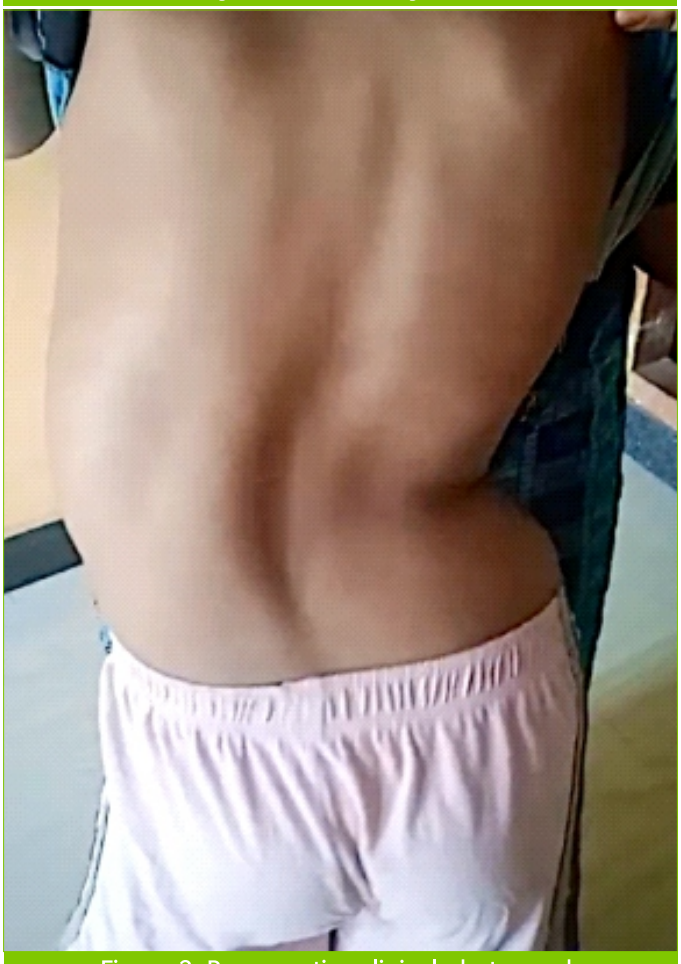

Figure 2: Preoperative clinical photograph depicting levoscoliotic deformity

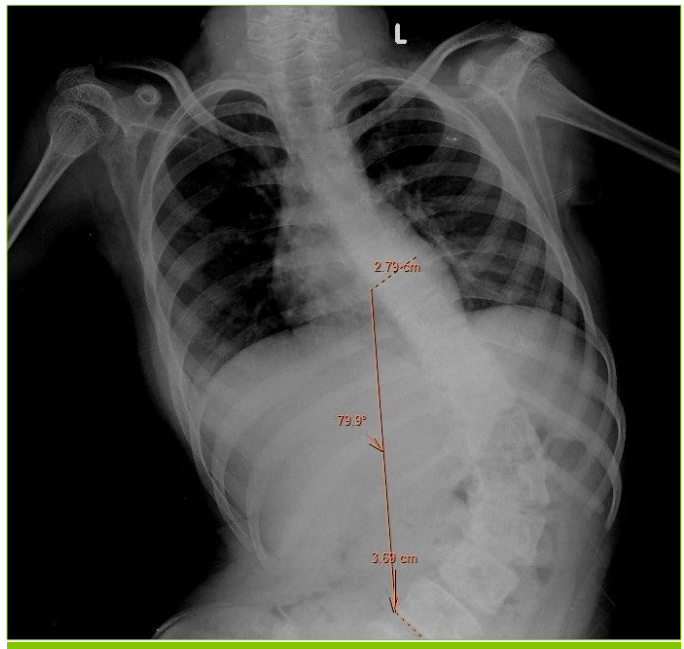

Figure 3: Xray with levoscoliotic curve with Cobbs angle measurement.

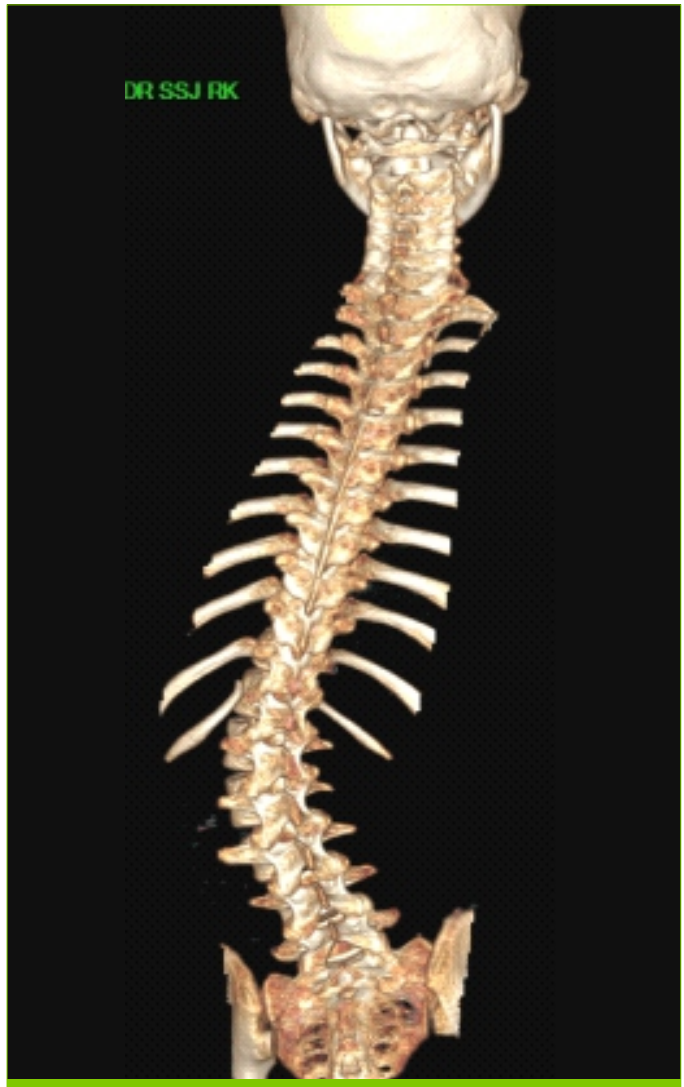

Figure 4: CT scan

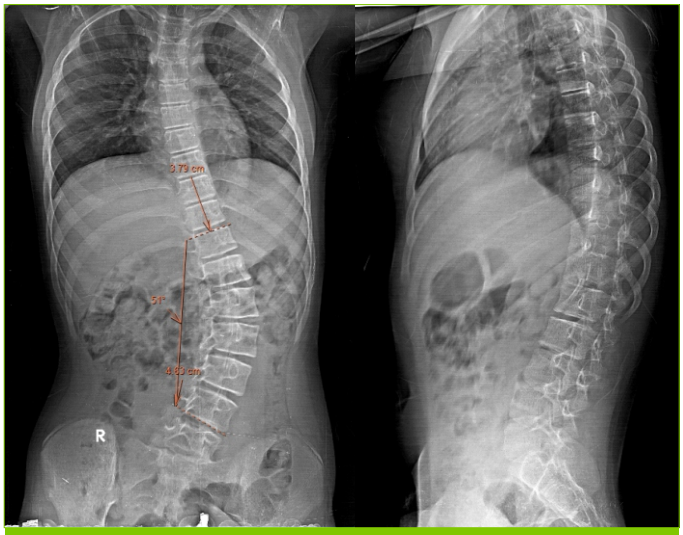

Figure 5: Traction Xray demonstrating partial correction of the curve.

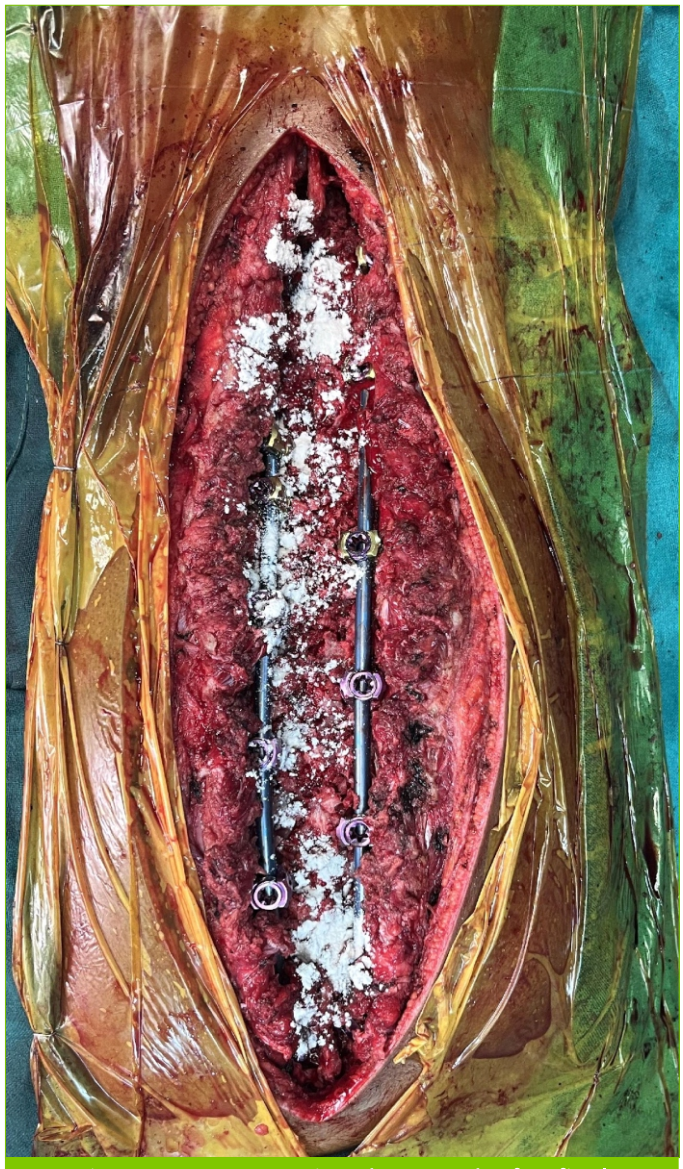

Figure 6: Intraoperative photograph after final instrumentation and deformity correction.

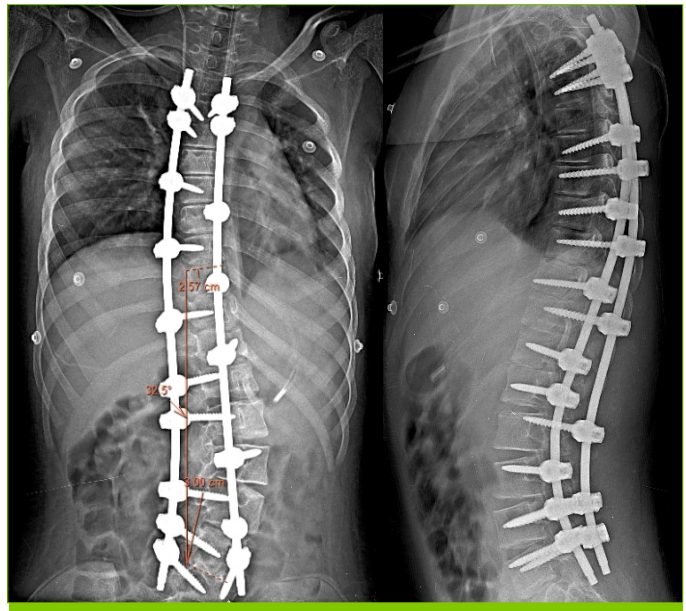

Figure 7: Postoperative Xray

Figure 8: Postoperative clinical photograph showing good coronal balance

DISCUSSION

Rett syndrome was first described by Andreas Rett in 1966. It is a progressive neurodevelopmental disorder caused by mutation of the MECP-2 gene (methyl CpG binding protein 2), located on the long arm of the X chromosome. It is a mostly sporadic sex-linked disease affecting approximately 1 in 10,000

females, being a common cause for mental retardation. Most affected males will not survive to term and those that will do will die by age 2 due to severe encephalopathy.

Four stages characterize the syndrome. The antenatal period is unremarkable and the child develops normally for approximately the first 6 months of life. The second stage is characterized by regression of acquired volitional hand movements and speech and occurs between 1.5 and 3 years of age. This regression stage is followed by a plateau phase where symptoms stabilize for several years. Finally, the late motor deterioration stage will result in epilepsy, progressive dystonia, rigidity and worsening ambulation. Spinal deformity is often observed in this phase.

The overall incidence of scoliosis in Rett syndrome is around 64%. The curves are usually treated when greater than 40°–50° to improve sitting balance, perceived pain and ease of activities of daily living. Patients with Rett syndrome often present considerable problems to the surgical team. These patients are typically underweight and undernourished which when combined with poor mobility results in osteopenia/osteoporosis with potential implications for the integrity of any bone/implant interface. Due to these reasons, literature suggests up to 100% of patients will experience some form of complications in the intra/postoperative period. Some reported complications in the literature are wound infections, pneumothorax, hemothorax, aspiration pneumonia, UTI, etc. Fortunately, our patient recovered uneventfully without any major complications.

CONCLUSION

Scoliotic deformity is common in patients with Rett syndrome and often will require surgical intervention to correct the deformity and to achieve acceptable coronal and sagittal balance. Rett syndrome patients have a high incidence of complications and due care has to be taken to prevent /reduce the occurrence of these complications.

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PRESURGICAL NASO-ALVEOLAR MOULDING (PNAM): THE CONCEPT, TECHNIQUE, CHALLENGES-AND THE EXPERIENCE @ SMILE TRAIN UNIT MHT

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INTRODUCTION

The cleft lip, alveolus and palate (CLAP) is one of the most common congenital malformations. In India, out of 24.5 million births per year, the birth prevalence of clefts is approximately 35,000.[1] These children may have cleft defects involving lip, nose, alveolus, hard palate and soft palate. These structures upon failing to fuse during embryogenesis give rise to cleft deformity. The degree of severity may vary depending on the structures involved, gap width, and the deficiency in the available tissue. The deformity is classified as unilateral when it affects one side of the face; however, when both sides are involved it is classified as a bilateral defect which is more complex to treat. Extensive deformity can present a significant surgical challenge in terms of achieving a good functional and aesthetic outcome.

Presurgical Infant Orthopaedics treatment was introduced in 1950 as a procedure to facilitate surgery and it has emerged as a valuable adjunctive therapy in the treatment of CLP. This is a nonsurgical orthopaedic treatment of moulding the infant bone to reduce the alveolar cleft width. It also comprises a nasal part to mould the nasal cartilages and a skin expansion component to augment the deficient tissue in the cleft deformity. Collectively the assembly is used with an aim of alleviating the asymmetry in the cleft deformity which facilitates more favourable primary lip, nose and palate repair. Thus the aforementioned correction not only enables better orientation of the alveolar bones, nasal cartilages and muscles after surgery but also reduces soft tissues closure in tension which reduces the scar formation post-surgery.

This paper provides a comprehensive review of the role of PNAM in CLP patients, discussing its mechanisms of action, sharing our experience on clinical efficacy and impact on surgical outcomes.

The Concept:

In 1984, Matsuo et al. postulated that neonatal cartilage is highly plastic and is amenable to manipulation due to the high levels of hyaluronic acid which is brought about by elevated levels of maternal estrogen circulating in the infant. This increased level of hyaluronic acid inhibits the intercellular matrix and enables relaxation of connective tissue, cartilages and ligaments and helps ease the passage of the fetus through the birth canal. Neonatal levels of maternally derived estrogen are maximal perinatally and fall to baseline levels in the subsequent 6- 8 weeks. Utilizing this window, Matsuo et al. was able to mould and correct deformed auricular cartilage; the same principle was then extended to the cleft nasal cartilage by Nakajima et al. and by Matsuo and Hirose. In 1993, Grayson et al. went a step further and combined the use of maxillary molding plates and nasal cartilage shaping, and described a technique to simultaneously correct the alveolus (with acrylic maxillary plates), lip (by soft- tissue taping) and nose (by using nasal stents to mold the highly pliable neonatal nose).

Mechanisms of Action of PNAM:

PNAM involves the use of custom-fabricated intraoral appliances to mold and reshape the alveolar segments, and surrounding soft tissues. By applying gentle pressure over the cleft segments, PNAM aims to gradually narrow the alveolar

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cleft width, improve nasal symmetry, and align the palatal shelves, thus facilitating primary lip and nose repair.

Clinical Efficacy of PNAM:

Numerous studies have demonstrated the clinical efficacy of PNAM in CLP patients. A systematic review by Maull et al. (2020) reported significant reductions in alveolar cleft width and nasal deformity following PNAM therapy, leading to improved surgical outcomes and aesthetic results. Additionally, PNAM has been shown to enhance maxillary arch alignment, improve feeding efficiency, and promote speech development in CLP patients.

Impact on Surgical Outcomes:

The integration of PNAM into the preoperative treatment protocol for CLP patients has been associated with several benefits. PNAM therapy allows for better approximation of cleft segments, resulting in improved tissue quality and vascularity, which facilitates primary surgical repair. Additionally, PNAM helps to minimize scar formation, reduce the need for secondary revisions, and improve long-term aesthetic outcomes following cleft lip and palate repair surgeries.

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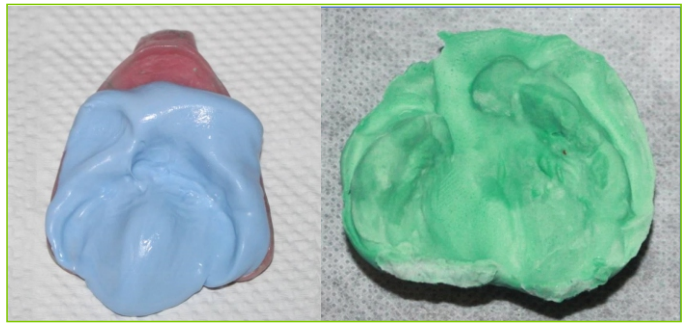
CLINICAL PROCEDURE:

Infant impression:

The primary impression of the patient with CLP is obtained within the 1st week after the birth. The rubber base impression material (Additional silicone) is used for recording the impression due to its ease of manipulation, good tear strength, controlled setting time, and accuracy. Customized impression trays are selected over stocked trays. Positioning of infant patients with CLP is of critical importance. Supine position with head down is the most preferred. The infants are fully awake without any anesthesia or premedication. Infants should be able to cry during the impression procedure and any absence of crying may indicate blockage of airway. Monitoring of infants' oxygen level is done throughout the impression recording procedure so as to prevent accidental hypoxia.

Appliance fabrication and design

The dental stone model is made using the impression taken. All the undercuts and the cleft space on the model are blocked out with the wax. The moulding plate is made up of hard, clear self-cure acrylic. A retention button is fabricated and positioned at an angle of 40° anteriorly to the plate. The orthodontic elastics and tapes should be used to secure the moulding plate adequately in the mouth. On the palatal surface of the moulding plate, a small opening of 6-8 mm diameter is made to provide an airway if the plate drops down posteriorly. The nasal hook is constructed and incorporated into the treatment when the cleft of the alveolus is reduced to about 5–6 mm in width.

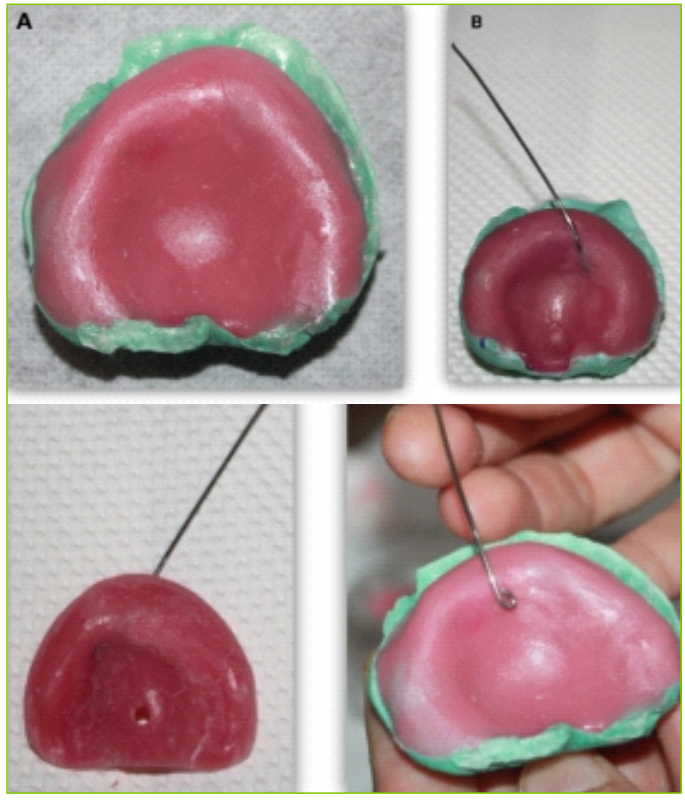


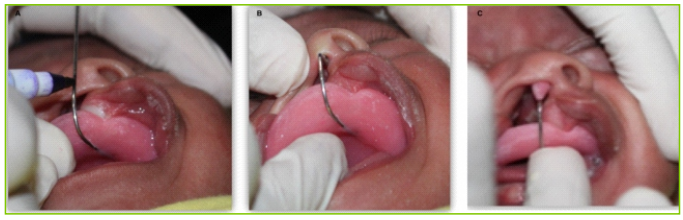
Moulding Technique

The adjustments are made by selectively adding the soft denture base material to the moulding plate and removing the hard acrylic. The moulding plate should not be modified more than 1 mm in one visit.

Lip taping and Nasal Hook

Extraoral tapes are used to bring the two lip segments together in conjunction with the moulding plate and nasal hook. Taping the lips improves the relation of lower mid-face skeletal as well as an improvement in the overlying soft tissue.





DISCUSSION:

The main objectives of the NAM technique have been cited as (i) repositioning maxillary segments in a favourable anatomical position (ii) facilitating primary lip, alveolar and nasal surgeries (iii) reducing nasal deformity (iv) improving the projection of nasal tip (v) facilitating feeding (vi) increasing the columella length and (vii) correcting septal position.

These corrections bring several benefits in the treatment of cleft lip and palate deformity. A proper alignment of the alveolus, lip and the nose helps the surgeon to achieve a better and more

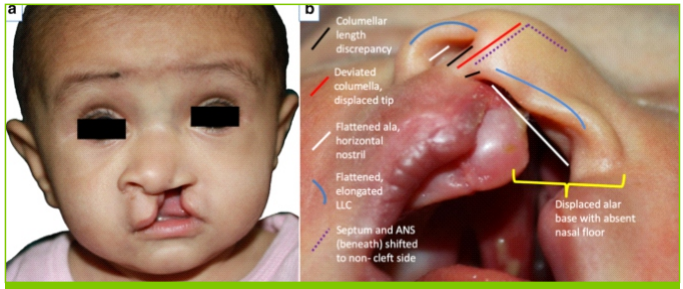


Photo courtesy: Krishnamurthy Bonanthaya, Jazna Jalil, Management of the Nasal Deformity in the Unilateral Cleft of the Lip and Nose, journal of Maxillofacial and Oral Surgery Volume 19, pages 332–341, (2020)

predictable surgical result. The cleft deformity is significantly reduced in size with the NAM therapy before surgery, making primary repair of the lip, alveolus and the nose more symmetrical. This improvement reduces the number of surgical revisions for excessive scar tissue, oronasal fistulas, and nasal and labial deformities.[3] With the alveolar segments in a better position and increased bony bridges across the cleft, the permanent teeth have a better chance of eruption in a good position with adequate periodontal support. In Bilateral CLP patients, an additional objective of centralization and retraction of the protruding premaxillary segment is achieved with the help of the appliance.[4]

Challenges and Considerations

Despite its proven efficacy, PNAM therapy poses several challenges and considerations. Firstly, the early reporting of the cleft infant is a key factor in the effective PNAM therapy. The small window period of neo-natal stage is crucial for effective moulding of the tissues in the desirable orientation and dimension. Therefore, one of the challenge is to bring awareness among the general practitioners especially paediatricians about the therapy and significance of early referral of the cleft infant to the cleft unit to make maximum out of PNAM. Secondly, the therapy demands significant clinical experience and expertise from the clinician for case selection, to appropriately guide bony movement, accommodate for the infant's growth and identify and alleviate undue pressure application. Moreover, trained teams who provide NAM are limited. The timing and duration of PNAM therapy must be carefully tailored to each patient's individual needs, taking into account factors such as cleft severity, skeletal maturity, and surgical readiness. Many such reasons have motivated cleft care providers to investigate the role of digital workflows in NAM therapy- to increase efficacy of treatment and enhance potential reach of care.Further, the patient compliance and caregiver involvement are crucial for the success of PNAM, as regular adjustments and monitoring are required throughout the treatment period.Most importantly, the care for this group of children is being delivered in a fragmented way through several centres in India. In fact there are no prospective randomised trials to evaluate the long term benefits of this procedure in Indian population. To address this gap a multicentric randomised controlled trial- the NAMUC study is being conducted and Smile Train unit at Meenakshi Hospital is one of the centres. This study is aimed to evaluate the

Photo: Anatomic landmarks illustrating the effective change brought after PNAM in a Unilateral Complete cleft lip palate deformity. Treated at Smile Train unit Meenakshi Hospital

Photo: Anatomic landmarks illustrating the effective change brought after PNAM in a Bilateral Complete cleft lip palate deformity. Treated at Smile Train unit Meenakshi Hospital.

effectiveness of the PNAM in patients with a non-syndromic complete unilateral cleft palate patients in the long term.

Our experience with PNAM cases

PNAM at our unit is started within 1 week of birth preferably to get the best compliance of the infant and the best results. It is continued till primary lip repair procedure. Customised plates are fabricated and delivered to the patient on their first visit. The plate not only serves to mold the alveolar bone in a desired contour but also serves as a barrier between the oral and the nasal cavity. This barrier simulates the palate which thereby provides a stable base against which the infant is able to suck bottle nipples. It also helps reduce the risk of aspiration as nasal regurgitation is minimised. Lip taping is also started simultaneously with the oral plate. When the cleft alveolar gap is reduced to an optimal width the nasal stent is incorporated in this assembly which enables the moulding of the nasal cartilages as well. Patient attenders are taught both the technique and the protocol of placing the plate and the stent in the baby's mouth. They are also educated about the protocol of hygiene, warning signs of pressure sores and other necessary precautions.

Conclusion

PNAM, when used prior to primary surgical lip repair should enhance surgical outcome, reduce the need for secondary dental, skeletal procedures and soft tissue revision surgeries in the future which thereby reduce the overall cost of treatment. It definitely facilitates infant feeding and nutrition which is crucial for growth and development of the newborn. A well-executed PNAM process by a committed team of Orthodontist and Surgeons supported by good compliance of the parents and child may prove to be a boon for a cleft lip palate child. Results of PNAM are promising for a child with CLAP, hence are encouraged to be used promptly after the birth and continued till further specific corrective surgeries are performed. Results of the ongoing randomised clinical trial at our unit will hopefully enrich us with stronger evidence to the current clinical practices.

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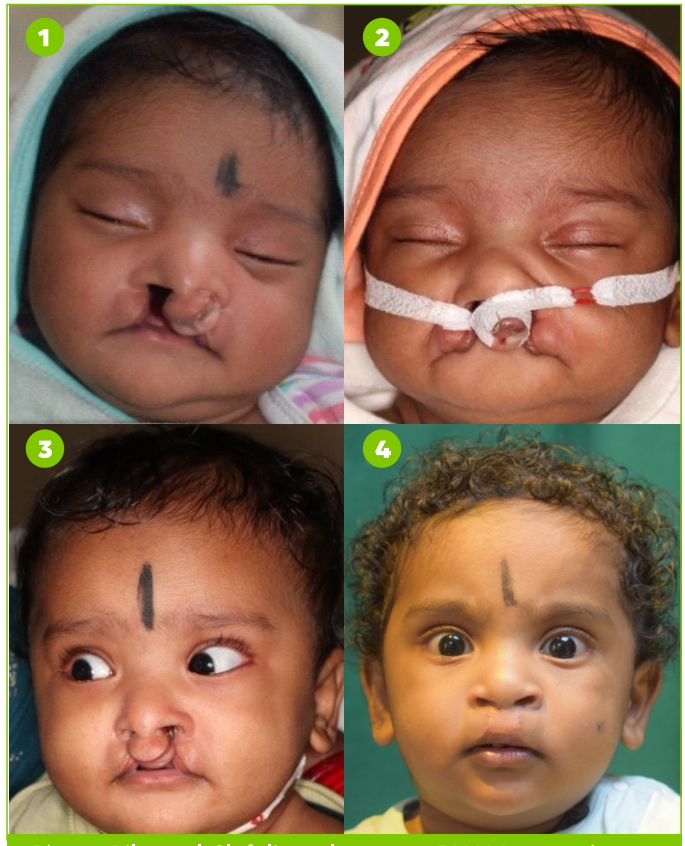

Photo: Bilateral Cleft lip palate case PNAM treated at Smile Train unit Meenakshi Hospital.

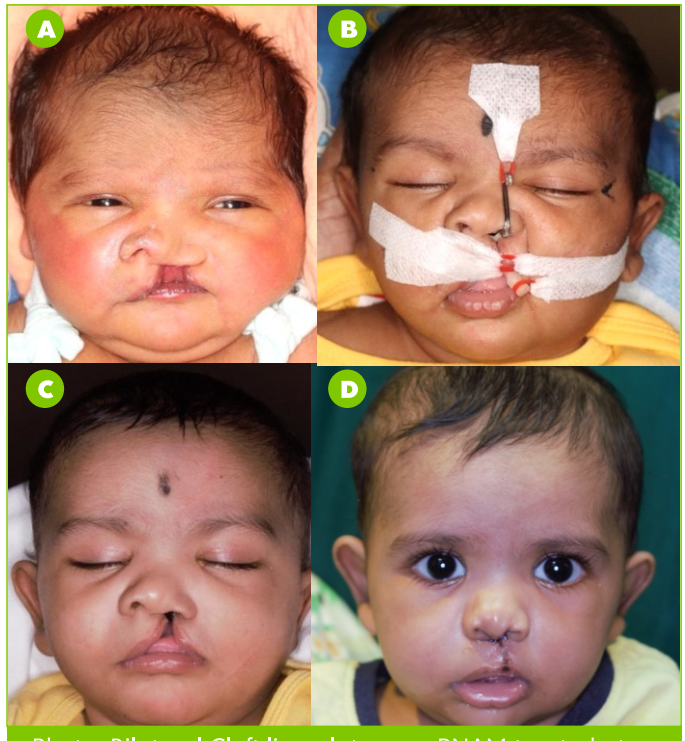

Photo: Bilateral Cleft lip palate case PNAM treated at Smile Train unit Meenakshi Hospital.

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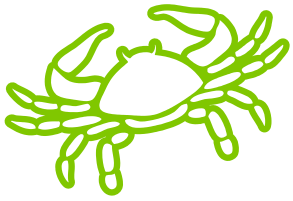

புற்றுநோய்க்கு முற்றுப்புள்ளி

புற்றுநோய்க்கு எதிரான போராட்டத்தில் நாம் எடுத்து வைக்கும் ஒவ்வொரு அடியும் வெற்றிதான்! நம்பிக்கையுடன் இருப்போம், புற்றுநோயை வெல்வோம்.

தஞ்சை மீனாட்சி மருத்துவமனையில் புற்றுநோய்க்கு எதிரான போராட்டத்தில், நாங்கள் உங்களுடன் இருக்கிறோம். புற்றுநோய் குறித்த விழிப்புணர்வை ஏற்படுத்தி, புற்றுநோயின் சவால்களுக்கு எதிராக உங்களுடன் ஒன்றிணைந்து நிற்கிறோம்.

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- அனுபவம் வாய்ந்த கதிரியக்க சிகிச்சை நிபுணர், புற்றுநோயியல் நிபுணர், புற்றுநோய் அறுவை சிகிச்சை நிபுணர் மற்றும் நோய் கண்டறியும் நிபுணர், ஆகியோரை உள்ளடக்கிய புற்றுநோய்க்கான சிகிச்சை குழு.
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