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THANJAVUR

MEDICAL JOURNAL

R N I .N o .T N E N G O 6 O 7 6





30.12.2023: New Year 2024 Grand Celebration event was successfully organized by Meenakshi Hospital in association with IMA Kumbakonam Branch. Chief Guest Dr.K.M.Abul Hasan State President,IMA-TNSB, Guest of Honor Dr.B.Sridhar State President, Elect 2025, IMA-TNSB and Dr.S. Veerapandiyan, Vice President, East Zone IMA, inaugurated the event. Dr. Kesavamoorthy, Senior Consultant and HOD, Cardiology, Meenakshi Hospital, Tanjore was felicitate during the function. IMA Kumbakonam Branch Office Bearers, President - Dr.S.Rajendran, Hony. Secretary - Dr.G. Sambasivam and Treasurer - Dr.S. Sujan, supervised the overall event. More than 150 Doctors participated with their families in the Gala event.

THANJAVUR MEENAKSHI MEDICAL JOURNAL

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FROM THE EDITOR'S DESK

Dear Readers,

Greetings and a Very Happy New Year.,

As we embrace the significance of Cervical Cancer Awareness Month (January), which emphasizes on creating awareness from screening innovations to treatment breakthroughs, Cervical cancer still remains a global health concern, but the strides made in recent years offer a glimmer of hope in the pursuit of better outcomes for affected individuals.

At the forefront of the battle against cervical cancer is the critical role of awareness and proactive screening. From the enduring importance of Pap smears to the advent of HPV testing, advocating for widespread awareness campaigns to encourage regular screenings are a continued necessity.

In the realm of treatment, personalized care is emerging as a beacon of progress. The latest advancements in radiotherapy and chemotherapy, emphasizes their role in tailoring interventions to the unique needs of each patient. From refined radiotherapy techniques to the promising landscape of targeted therapies, these innovations are reshaping the treatment landscape.

This month serves as a reminder that the fight against cervical cancer extends beyond awareness and screenings—it extends to ensuring that groundbreaking treatments reach every corner of our communities.

Beyond the pages of research and clinical updates, it is the collective voice of advocates, survivors, and healthcare professionals that propels the message of cervical health into the public consciousness. We appreciate the tireless efforts of those working towards destigmatizing discussions around cervical health and fostering a culture of proactive care.

In conclusion, we as doctors and as a beacon of awareness and knowledge during Cervical Cancer Awareness Month, let us continue to amplify the conversation, break down barriers to screening, and advocate for a future where cervical health is prioritized and protected.

Wishing you an enlightening and impactful read,

'COMMON CANCERS ARE PREVENTABLE AND CURABLE'

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SPOTTER - 01



Quiz Details:

Identify the Condition

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





Quiz prepared by

Dr. P. Thendral, MBBS, DO, MS (Ophthal), Fellowship in CRS. Sr.Consultant - Ophthalmology ,
Meenakshi Hospital

- Send your answer within 20th MARCH 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 - 17



Quiz Details:

Risk of severe mental retardation by prenatal radiation exposure is maximum during:

- A). 8-15 weeks post conception
- B). Preimplantation period
- C). 16-25 weeks post conception
- D). Last trimester

Answer: A). 8-15 weeks post conception

last Issue





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GUNSTOCK ELBOW DEFORMITY – CORRECTIVE OSTEOTOMY – CASE SERIES



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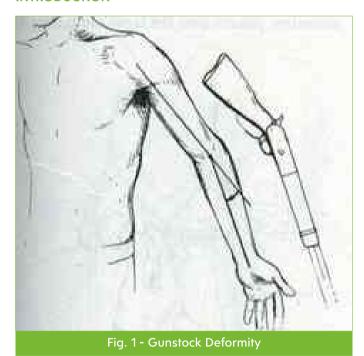
Associate Consultant

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Meenakshi Hospital

ABSTRACT: Gunstock elbow deformity is the most common post traumatic elbow deformity occurring in pediatrics. This neglected elbow deformity worsens with bone growth causing functional dysfunction, cosmetic disfigurement and psychological impact to children. We have corrected nearly 20 children with gunstock deformity using lateral closing wedge corrective osteotomy surgeries and had given a good functional outcome.

INTRODUCTION

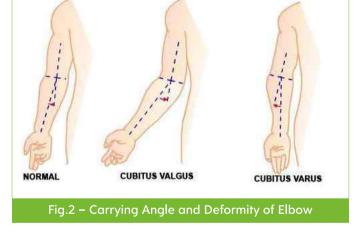


Cubitus varus deformity is also known as Gunstock deformity since it looks like loading stock of old long barrel guns.

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Carrying angle of elbow is normally the valgus angle between the long axis of arm and extended forearm, usually varies between 5 to 15 *. Cubitus valgus deformity is the increased valgus angle of elbow and Cubitus varus deformity is the reduced valgus angle of elbow. Cubitus varus is the most common angular deformity that results from supracondylar fracture as late complication in skeletally immature children between 5 to 10 years of age. Post traumatic cubitus varus may predispose a child to subsequent lateral condylar fracture also. Causes for cubitus varus deformity are medial displacement,

Causes for cubitus varus deformity are medial displacement, rotation and varus tilting of distal fragments. Malrotation of distal humerus is compensated for to a large degree by motion of the shoulder joint.



Fig.3 – Right Elbow Cubitus Varus Deformity



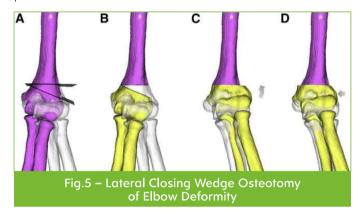
Fig.4 - Left Elbow Cubitus Varus Deformity

Three basic types of osteotomies

- -Medial opening wedge osteotomy with bone graft
- -Oblique osteotomy with derotation
- -Lateral closing wedge osteotomy

Three-dimensional osteotomy for correction of cubitus varus deformity in which medial, posterior tilt and rotation of distal fragments can be corrected.

Lateral closing wedge osteotomy is the easiest, safest and the most stable osteotomy. Methods of fixation are use of 2 screws with a wire, plate fixation, crossed K wires and staples. Supracondylar osteotomy for cubitus varus is a reconstructive procedure.



Lateral Closing Wedge Osteotomy

Tourniquet inflated, lateral incision, under fluoroscopic quidance, insert 2 K wires into lateral condyle before osteotomy and advance them just distal to the planned distal cut. Closing wedge osteotomy laterally leaving the medial cortex intact. Weaken the medial cortex using drill holes and apply a forceful valgus stress to complete the osteotomy with the forearm in pronation and elbow flexed. Close the osteotomy and advance the K wires from the lateral condyle into the medial cortex of the proximal fragment.



Fig.6 – C arm images of Wedge Osteotomy Correction



Fig.7 – Operative picture of Lateral Wedge Osteotomy

Splint the arm in 90* flexion and full pronation. Wires are removed approximately 6 weeks after surgery and range of movements started. Patients were reviewed periodically for physiotherapy care and functional outcomes were assessed. Complications which we have encountered are transient joint stiffness, delayed osteotomy union and minimal persistent deformity of elbow.



Fig.8 - Corrected Deformity of Right Elbow in 19 years Adolescent



Fig.9 - Corrected Deformity of Right Elbow in 8 years Girl



Fig.10 – Functional Outcome of 8 years Girl with Right **Elbow Deformity Correction**



Fig.11 – Functional Outcome of 5 years girl with Right **Elbow Deformity Correction**



Fig.12 – Functional Outcome of 12 years Girl with Right Elbow Deformity Correction

CONCLUSION

Lateral closing wedge osteotomy for the correction of cubitus varus deformity of the elbow is the safe and effective method which gives near normal elbow with minor complications and cosmetically more acceptable to parents.

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SYLVIAN FISSURE EPIDERMOID CYST - A CASE REPORT



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ABSTRACT: Epidermoid tumors are benign tumors which contain keratin, cellular debris, and cholesterol, and are lined with stratified squamous epithelium. They grow in discreet silence sustained over a multitude of years. The tumors most commonly present with headache and seizures. We report the case of a 53-year-old male with a large sylvian fissure epidermoid tumor who presented with seizure. The patient was operated and total excision of the tumor was performed.

INTRODUCTION

Epidermoid tumors comprise about 0.2–1% of all primary intracranial tumors. These tumors are commonly located in the cerebellopontine angle, in the cisterns around the tentorium and in the suprasellar and parasellar cisterns.[1,2] Occasionally, these lesions occur in the Meckel's cave, in the middle fossa, diploe of bone, and in the spinal canal.[1,2,3],sylvian fissure .Symptoms are usually related to the position of the cyst. The most frequent symptoms are headache and seizures.[1] Uncommonly, they may present with chemical meningitis or psychosis. We present a 53-year-old man with a sylvian fissure epidermoid cyst .Our literature search revealed only a few cases of a sylvian epidermoid tumor were reported so far.

CASE REPORT

A right-handed 53-year-old gentleman presented to us with GTCS for the last 3 months. He had occasional headache for past 2 yrs. His neurological examination was normal. Magnetic resonance imaging (MRI) of the brain showed an irregular lesion in the left sylvian fissure with extensions in the frontal and temporal operculum. The lesion was hypointense on T1-weighted imaging, (Fig. 1a) hyperintense on T2-weighted

imaging with (Fig 1b)no enhancement on contrast. The lesion showed restricted diffusion(Fig 1c) .There was encasement of the middle cerebral artery. A diagnosis of left sylvian fissure epidermoid tumor was made, and the patient was operated. A left frontotemporal craniotomy was performed and the dura was opened based on the sphenoid ridge. The sylvian fissure was splayed open by the pearly white epidermoid tumor (Fig 2a). A tumor debulking began. The tumor had encased the middle cerebral artery and its branches(Fig 2b). A total excision of the tumor was performed. (Fig 2c) At the end of the surgery, a panoramic view of middle cerebral artery was seen. The patient was well following the surgery. At a follow-up of 1-year, the patient is well and is back to all his routine activities. Tumor was pearly white in colour and Histopathology of the tumor revealed the characteristic features of an epidermoid tumor.

Preoperative images



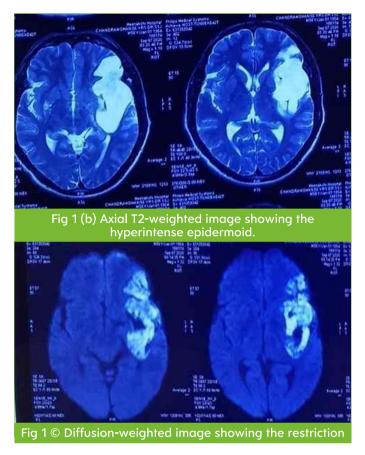
Preoperative images of a 53 -year-old male with left sylvian fissure epidermoid cyst.

CORRESPONDING AUTHOR

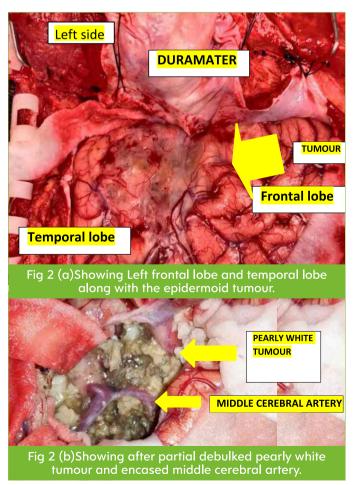
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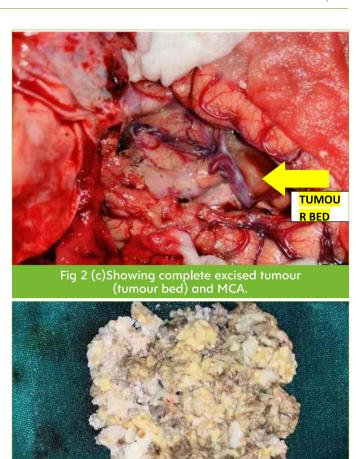
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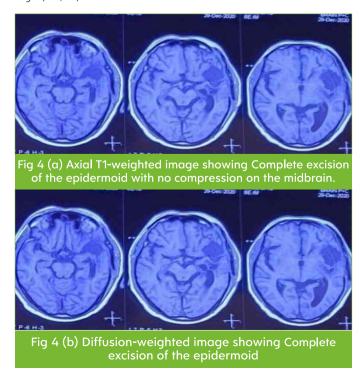
INTRA OPERATIVE IMAGES





Postoperative MRI Showed Completexcision of Tumor Fig4 (4a,4b)

Fig 3: Post op specimen.



DISCUSSION

Epidermoids are slow-growing benign tumors and have a peak incidence in the fourth decade. Typically patients are between 20 and 40 years of age. This so-called "beautiful tumor" has an irregular cauliflower-like outer surface that grows and encases vessels and nerves. Due to their slow growth, they develop a symbiotic relation with the brain and do not cause many neurological deficits.[3] Signs and symptoms of epidermoid cysts are due to gradual mass effect, with presentation including: Headaches (most common), cranial nerve deficits, cerebellar symptoms, seizures, raised intracranial pressure.[1,2] Recurrent aseptic meningitis is uncommon but recognized similar to the less common dermoid cyst.[13] 1] Occasionally, cerebellopontine angle epidermoid tumors can present with trigeminal neuralgia and tic convulsion[6]

EMBRYOLOGY

•They may arise from displaced dorsal midline ectodermal cell rests between the third and fifth weeks of embryogenesis during neural tube closure.

The usual locations of epidermoid cysts are the parasellar region and cerebellopontine angle, and it is less commonly located in suprasellar region, cerebral and cerebellar hemispheres, sylvian fissure, and lateral and fourth ventricles. Epidermoid cysts located in the posterior fossa usually arise in the lateral subarachnoid cisterns, and they are rarely located in the brain stem. [4,5].

The major differential diagnosis for an epidermoid cyst are arachnoid cysts, hamartomatous lipomas, dermoid cysts, cystic neoplasms, [7,8] neurocysticercosis, neurenteric cyst etc.

They can usually be differentiated by CT scan or MRI images of the brain. Conventional MR images sometimes cannot reliably be used to distinguish epidermoid tumors from arachnoid cysts since both lesions are very hypointense relative to brain parenchyma on T1-weighted MR images and very hyperintense on T2- weighted images. In contrary, fluid-attenuated inversion recovery (FLAIR) and diffusion-weighted (DW) sequences can successfully be used for diagnosis of epidermoid cysts by revealing its solid nature.[10-14] DW imaging is superior to other MR sequences in delineating the borders of the epidermoid cyst. FLAIR MR imaging is based on the nulling of the signal from CSF. Epidermoids characteristically present on this sequence as heterogeneous lesions with central parts of the tumor being hyperintense relative to the hypointense CSF.

The surgical approach is generally determined by the location and the extent of the lesion.

•Surgical excision is the treatment of choice if symptomatic. However, complete resection is difficult as not all tissue can be removed, especially from around cranial nerves and vessels. Recurrence is therefore not uncommon, although growth is typically slow and many years can elapse without new symptoms. Malignant transformation of an epidermoid cyst into a squamous cell carcinoma is a known but extremely rare occurrence. Malianant transformation is suggested by enhancement after contrast administration or rapid growth.[4]

•Chemical meningitis can occur by spillage of the cyst contents during operation, which usually is transient and self-limiting[9] and can be managed successfully with steroids

In the management of such tumors, one should keep in mind that an aggressive radical surgery carrying a high morbidity and mortality and a conservative subtotal tumor excision is associated with a higher rate of recurrence, but early diagnosis and complete excision or near total excision of this benian tumor can cure the patient with the expectation of normal life.

CONCLUSION

Sylvan epidermoid is one of the rare sites for epidermoid location .MRI DWI is an important diagnostic image. Complete excision is challenging one and can cure the patient with the expectation of normal life.

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ANOTHER FEATHER IN THE CAP OF OUR GREAT PATIENT CARE AHPI "HOSPITAL NON CLINICAL OPERATIONS

EXCELLENCE AWARD"





THIS RECOGNITION REASSURED OUR COMMITMENT OF "PATIENT CENTRIC APPROACH"

TO THE PEOPLE IN DELTA REGION



DR. M. KARTHIKEYAN MBBS, MS, DNB(ORTHO), MRCSED, Fellowship in Spine Surgery (Australia, Denmark & India) RP - NUH (Singapore) Senior Consultant - Department of Orthopaedics & Spine Surgery Meenakshi Hospital

INTRODUCTION

Vertebral hemangiomas (VH) are benign vascular neoplasms often found incidentally in the vertebral column. It commonly involves the thoracic and lumbar spine, with the peak incidence in the fourth to sixth decade. They are the most common vertebral tumors. VHs are characterized by the presence of multiple thin-walled newly formed vessels surrounded by the fat with infiltration of the medullary cavity between bony trabeculae. Though most are asymptomatic, 0.9% to 1.2% of patients become symptomatic and are defined as aggressive when there is extension into the spinal canal leading to spinal cord compression and myelopathy. Cord compression is often progressive but may occur acutely in some cases. It is still not clear why certain hemangiomas turn aggressive and become symptomatic while most follow a benign course.

Different modalities have been described for AVHs like laminectomy decompression, vertebroplasty, en bloc spondylectomy, embolisation and radiation therapy. Surgery decompresses the spinal stenosis with a small possibility of bleeding-induced stenosis. This risk can be mitigated by preoperative embolisation, however, this facility is not universally available in all hospitals.

CASE REPORT

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hemangioma in a 60-year-old female with thoracic

In this case report, I describe a case of aggressive vertebral

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myelopathy. She presented with non-specific upper and mid back pain for a duration of 1 year. 2 months before the presentation, she also noticed thoracic band-like pain associated with unsteadiness while walking. There was also subjective weakness in both lower limbs and paresthesias in both lower limbs. She presented to us when she started developing symptoms of urinary incontinence for 2 weeks. On examination, she had deep tenderness in her upper back pain. Neurological examination demonstrated spasticity of both lower limbs with hyperreflexia and clonus. She had a 3/5 power and altered sensation in her lower limbs. Tandem gait was not possible and Romberg's sign was positive. Plantars were upgoing bilaterally. Clinical examination was suggestive of thoracic myelopathy. She was admitted and radiological investigations were performed. MRI (plain and contrast) demonstrated a lesion in the T2 vertebra with altered signal intensity in both T1 and T2 encroaching into the canal with



severe cord compression (Figure 1&2). CT sagittal and axial cuts showed a "polka dot appearance" of the T2 vertebra with

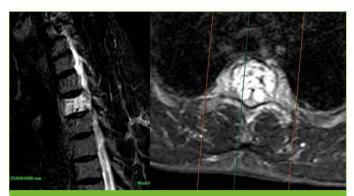


Figure 2: Preoperative contrast MRI showing extension of the lesion into the spinal canal and no involvement of posterior element

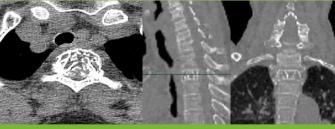


Figure 3: Preoperative CT demonstrating classical polka dot appearance of vertebral hemangioma

the extension of soft tissue with calcification into the spinal canal. There was no involvement of posterior elements (Figure 3). The patient was counseled for surgery. After the surgical workup, she underwent a T2 wide laminectomy, left transpedicular vertebroplasty and T1 to T3 posterior



Figure 4: Postoperative Xray with T2 vertebroplasty and T1 to T3 pedicle screw instrumentation

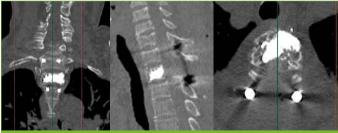


Fig5: Postoperative CT scan demonstrating the satisfactory cement distribution in the lesion and decompression.

instrumentation and fusion (Figures 4 & 5). Intraoperatively no complications were encountered and special attention was paid to minimize blood loss. Postoperatively, she had noticed an improvement in lower limb paresthesias and thoracic bandlike pain. Her lower limb power gradually improved during the postoperative period. At 6 weeks follow-up, she can walk with the help of a walker. She has regained control of her bladder habits.

DISCUSSION

Vertebral Hemangioma (VH) can be of three types based on the symptoms and they are asymptomatic VH, painful VH, and compressive spinal hemangioma. VHs become aggressive with the resorption of the underlying bone, leading to pathological compression fracture and then neurological deficit. The mechanism of AVHs causing vertebral pathological fracture is that hemangioma might cause diffuse bone infiltration and weaken the bone mineral density. Enlargement of the affected vertebrae and the compression fracture can cause spinal canal narrowing and spinal cord compression. The diagnosis and treatment of these AVH are challenging.

In this patient, vertebroplasty was performed in addition to decompression. Injection of polymethylmethacrylate(PMMA) cement into diseased vertebral bodies is vertebroplasty and it causes an irreversible hardening of the hemangiomatous vessels, thus obtaining an antalgic effect and vertebral stabilization. One has to be aware of complications like cement leakage into the spinal canal resulting in paraparesis, cement embolisation and massive blood loss especially when attempting to do a corpectomy. No such complications were encountered in our patient.

CONCLUSION

The treatment of AVHs with neurological deficits is challenging and a proper understanding and execution of different treatment options is of paramount importance to provide a satisfactory outcome to these patients.

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TRANSFACET SCREW FIXATION OF THE SUBAXIAL CERVICAL SPINE—HOW I DO IT?



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ABSTRACT: Background Transfacet screw fixation is a biomechanically effective way of fusing the subaxial cervical spine. The technique used by this author is Muthukumar's technique.

Methods With the patient in prone position, a conventional posterior exposure of the cervical spine is done. The entry point used by this author is 2 mm above the middle of the lateral mass without any lateral angulation. Under fluoroscopic auidance the facet is drilled until all the four cortical surfaces are purchased. Then the depth is measured to assess the length of the screw required. This is followed by tapping and screw insertion both of which are done under fluoroscopic control. All screws are placed prior to laminectomy to decompress the cervical cord.

Conclusions This is a simple, inexpensive and biomechanically effective way of stabilizing the subaxial cervical spine.

Key Words: Cervical spine.Laminectomy.Posterior instrumentation.Posterior decompression .Spinal fusion . Transfacet screw fixation

RELEVANT SURGICAL ANATOMY

The lateral masses of the cervical spine are composed of the inferior articular process of the vertebra above and the superior articular process of the vertebra below. In the cervical spine, because of the orientation of the facets, the lateral masses are in the same plane as the lamina. Hence, identifying the centre of the lateral masses is crucial as the entry point is based on this point. There are two structures at risk during this procedure, viz, the cervical nerve root and the vertebral artery [3]. However, with proper surgical technique injuries to these neurovascular structures can be avoided.

DESCRIPTION OF THE TECHNIQUE

This procedure is done with the patient in prone position. During exposure, the neck is placed in neutral position. A conventional midline exposure of the posterior aspect of the cervical spine is done, making sure that the boundaries of all the lateral masses are adequately exposed. In the literature, there are two methods that are described for transfacet screw

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fixation of the subaxial cervical spine: the Takayasu technique and the Dalcanto technique [5, 6]. However, this author uses a different technique, with a more rostral entry point, as both the above techniques are associated with unacceptable rates of fractures of the facets. In this author's technique, the entry point is situated 2 mm above the middle of the lateral mass. To identify the entry point, all the boundaries of the lateral mass should be adequately exposed. The facet joint is then curetted to facilitate fusion. If necessary, bone harvested from the spinous processes can be packed into the joint [5]. This is followed by preparing the entry point with a small 2 mm burr to avoid skiving of the drill bit which can result in fracture of the lateral mass while drilling. After entry site preparation, the facet is drilled manually under fluoroscopic control, so that the trajectory is perpendicular to the facet joint, until all the four cortical surfaces of the facet joint are crossed [1, 2, 4]. In this author's technique, there is no lateral angulation as described by Dalcanto. The surgeon can "feel" the passage of the drill through the four cortical surfaces. Once the drill passes through all the four cortical surfaces as confirmed by fluoroscopy, then this is followed by measuring the length of the screw required by using a K wire. Subsequently, the drilled hole is tapped with an appropriate sized tap until all the four cortical surfaces are

purchased as visualized by fluoroscopy. This is followed by the placement of a 3.5-mm screw of appropriate length. In this author's experience, most patients require a screw with a length of 16 mm. Occasionally, depending on individual patient's anatomical variations, some facets may require screws with lengths of 14 mm or 18 mm . As this author's technique uses an entry point 2 mm above the middle of the lateral mass, the length of the screws required will be slightly longer than while using the Takayasu's or Dalcanto's techniques. The same procedure is then repeated at the all the required levels. It is important to ensure that the cervical lordosis is restored prior to placement of screws. Restoration of lordosis is done by appropriate positioning of the cervical spine under fluoroscopic control prior to screw hole preparation. Laminectomy is generally done after screw insertion so that accidental slippage of instruments does not harm the cervical cord. After laminectomy, the bone acquired by laminectomy can be placed over the decorticated lateral masses to facilitate fusion. Screws can also be placed in levels adjacent to laminectomy if these levels exhibit instability. The above described technique is suitable for inserting transfacet screws from C3 to C6 levels (Figs. 1, 2, 3 and 5) Postoperative CT Cervical Spine Show four Critical purchase of trenofacetel show (4a,4b). The facets at C7 level are transitional between cervical and thoracic anatomy and are usually thin and elongated, and

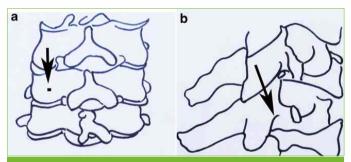


Fig. 1 Diagrammatic representation of the entry point and the The entry point is located 2 mm above the middle of the lateral black dot (a). There is no lateral angulation as shown by the arrow (a). The screw should be perpendicular to the facet joint and should purchase all the four cortical

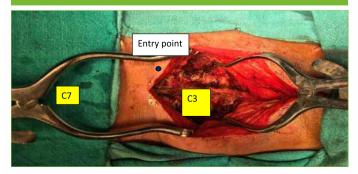


Fig .2 Intra-operative photograph showing the drill in the entry point located 2 mm above the middle of the lateral mass. Note: there is no lateral angulation involved



Fig 3 Intra-operative photograph showing the drill in the entry Note: there is no lateral angulation involved



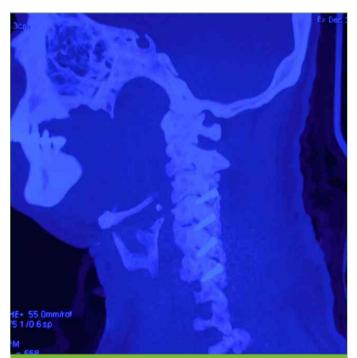
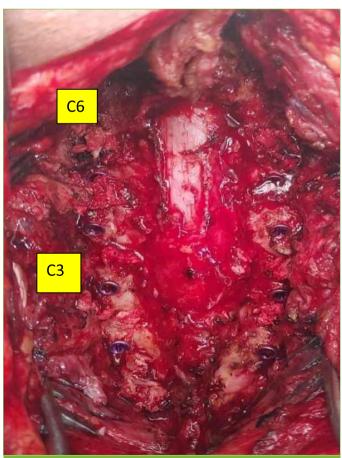


Fig 4 b) Sagittal reconstructed CT scans showing the screws across the facet joints purchasing the four cortical surfaces of the facet joints of both sides



5 Intraoperative photographs showing screws placed in -C4, C4–C5, C5– C6, C6–C7 in a before laminectomy and b after completion of laminectomy

both sides can be studied to find out the feasibility of this technique in individual patients. 2. Fracture of the facets that is known to occur with both Takayasu and Dalcanto techniques can be reduced, though not completely eliminated, by using the entry point described by this author.

SPECIFIC PREOPERATIVE AND INTRAOPERATIVE CONSIDERATIONS

- 1. Fixed cervical kyphosis as determined by preoperative dynamic radiography is an absolute contraindication for this procedure.
- 2. Fracture of the facets can occur in elderly patients with osteoporosis and hence, special care should be taken in these patients while drilling and tapping.
- 3. It is important to restore the cervical lordosis prior to placing the instrumentation.

POSTOPERATIVE CONSIDERATIONS

Patients are usually advised to wear a hard cervical collar for 3 months postoperatively.

Information to be given to the patient about the procedure and the risks

Patient should be counseled that there is likely to be some restriction of neck movements and they should wear a hard cervical collar for 3 months. They should also be counseled about the small risk of cervical nerve root injury and vertebral artery injury even though this author has not encountered both so far.

KEY POINTS

- 1. Transfacet screw fixation of the subaxial cervical spine is an inexpensive technique for stabilizing the subaxial cervical spine. As conventional screws are used instead of specialized lateral mass screws, this technique is inexpensive [4].
- 2. This technique is biomechanically equivalent or even superior to lateral mass fusion, as in this technique four cortical surfaces of the facets are purchased by the screws instead of two cortical surfaces as in lateral mass fusion [2].
- 3. Entry point for the screws should be 2 mm above the middle of the lateral mass in the technique used by this author.
- 4. Screw should be perpendicular to the facet joint.
- 5. There should be no lateral angulation.
- 6. Preparing the entry point with a 2-mm burr or an awl reduces the incidence of fracture of the facets.
- 7. Using the above technique, most patients require screws with lengths of 16 mm. Occasionally, 14 mm or 18 mm screws may be necessary depending on the patient anatomy and level.

- 8. The diameter of the screw is 3.5 mm.
- 9. It is important to restore the cervical lordosis prior to screw placement.
- 10. Care is required while drilling and tapping to avoid fracture of the articular processes.

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A CASE OF PYCNODYSOSTOSIS - REVIEW OF CLASSICAL **IMAGING FINDINGS WITH CRANIOSYNOSTOSIS** - A RARE ASSOCIATION.



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ABSTRACT: Pycnodysostosis is a rare skeletal dysplasia, which includes disproportionate short stature, craniofacial abnormalities such as frontal and occipital bossing, small face, beaked nose, hypoplastic mandible, abnormal dentition and hypoplastic terminal fingers and toes. In addition to these, hypoplastic lateral end of clavicle, diffuse bone sclerosis and segmentation anomalies of spine are also reported. Although classical clinical findings may be present, imaging plays an important role in the diagnosis of this condition by ruling out other osteosclerotic skeletal dysplasias.

INTRODUCTION

Pycnodysostosis is a rare autosomal recessive skeletal dysplasia, which due to deficient activity of lysosomal protease cathepsin K gene. The disease has an equal sex distribution with an incidence of 1-1.7 per million births (1). Patients with this condition typically have disproportionate short stature, craniofacial abnormalities such as frontal and occipital bossing, hypoplastic mandible, high arched palate and abnormal dentition. The hands and feet are small with hypoplastic terminal fingers and toes. Osteosclerosis is more obvious in long bones with delayed closure of cranial sutures. Here we present the classical imaging findings of this rare skeletal dysplasia in a 10-year-old boy.

CASE REPORT

A 10-years old boy born of consanguineous marriage presented with complaints of headache to pediatric OPD. On general examination the patient height was -103cms (height for age: < 3rd percentile and Z-score was -3). The weight of the patient was 20kgs (weight for age: < 3rd percentile and Z-score was -3) and head circumference was 48cms.On facial examination, there was frontal and occipital bossing with depressed mid face and maxillary hypoplasia. Hard palate showed furrows like depression for the entire length. Soft palate appeared normal. On Dental examination, mixed and delayed dentition for the age was noted. Brachydactyly was



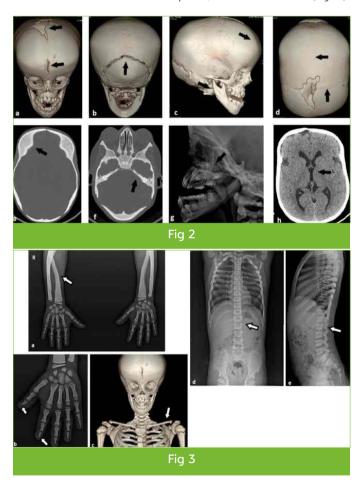
noted in hands and feet (fig 1). Neurological examination was normal without any previous history of developmental delay.

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His school performance was normal as stated by his parents. IQ tests done elsewhere showed average IQ score (90-109: WAIS-III). Other systemic examinations were unremarkable. Family history of similar appearance was noted in late paternal grandfather for which medical records were not available. No evidence of similar abnormality noted in his elder brother. Routine blood investigations were done. All haematological parameters were within normal limits with Haemoglobin – (11.5) gm/dl) and renal parameters were normal. Based on the clinical imaging findings skeletal dysplasia was considered and the patient was referred to radiology for further evaluation.

On Radiological examination, CT Brain showed diffuse increase in skull bone density - s/o Osteosclerosis (fig 2).



Delayed closure of cranial sutures with open anterior fontanelle, lambdoid and metopic sutures. Wormian bones were absent in this case (fig 2). Sagittal and coronal sutures craniosynostosis with frontal and occipital bossing noted (fig 2). Brain parenchyma was normal. No evidence of hydrocephalus or mass lesions in brain parenchyma (fig 2). Hence headache was attributed to craniosynostosis, which is an extremely rare association. Obtuse mandibular gonial angle with relative prognathism and hypoplastic maxilla with crowded tooth sockets noted (fig 2).

X-ray AP view of both hands showed mild medullary expansion of distal radius with remodelled and narrow terminal phalanges, simulating acro-osteolysis (fig 3). X-ray AP and lateral view of the dorso-lumbar spine showed increased bone density. No segmentation anomalies were present (fig 3). Normal appearance of clavicle without aplasia/hypoplasia (fia3).

DISCUSSION

Pycnodysostosis is a rare autosomal recessive skeletal dysplasia syndrome. It is due to deficient activity of lysosomal protease cathepsin K (CTSK) gene, which degrades collagen and osteonectin in osteoclasts. 33 Mutations which includes nonsense, missense and stop codon mutations in the cathepsin K gene, on Chromosome 1q21 have been reported (2). Deficient activity of CTSK gene is manifested clinically as osteosclerosis, which leads to increased bone density and stress fractures in weight bearing long bones (3).

The clinical profile of the patient includes disproportionate short stature, craniofacial deformity, prognathic jaw, short and stubby fingers / feet (6). The intra-oral features include groove in hard palate, overcrowded/persistent primary teeth and high arched palate (5).

The radiological findings include failure of cranial sutures to close with a wide anterior fontanelle and numerous wormian bones, predominantly in the lambdoidal region (4). Frontal and occipital bossing are present. Craniosynostosis may be seen in association with pycnodysostosis but is extremely rare, as there are only a few cases described in literature (7). Obtuse mandibular gonial angle with relative prognathism and hypoplastic maxilla are typical facial features (4). Acroosteolysis of distal phalanges of fingers and toes, abnormal dentition, hypoplastic clavicle & mandible are also seen. Spine anomalies include hyper-lordosis, scoliosis, kyphosis and block vertebrae (5).

In some cases, madelung's deformity, spoon shaped nails, spool shaped vertebrae, spondylolysis of C2 and spondylolisthesis of lower lumbar vertebrae, fractures of clavicle and mandible, deformed rib cage with pectus excavatum are also reported. Shallow and obliquely roofed acetabula and coxa valga may be seen in the pelvis (8).

Other systemic manifestations of this autosomal recessive condition includes fragile bones, which might lead to increased risk of fractures. Delayed fracture healing can be a complication as there is incomplete remodelling associated with osteoclast dysfunction. Stridor and laryngomalacia are seen in 20% patients which might lead to obstructive sleep apnea syndrome. Ocular abnormalities include refractive

disorders and strabismus, though blindness can occur in association with raised ICT in few cases. Chest deformities (Kyphosis, narrow chest) can also be seen though in very few cases. Intelligence is typically normal in affected patients, except when associated with brain malformations (9).

Anaemia and splenomegaly are not the features of Pycnodysostosis, as medullary canal is preserved in this condition (10.11). Histologically normal cortical bone with decreased osteoblastic and osteoclastic activity are noted. The lamellar bone is sclerotic and contains a large amount of woven bone with a mosaic pattern. Howship's lacunae are infrequently seen (12).

The differential diagnosis for Pycnodysostosis include, osteopetrosis, cleidocranial dysplasia, melorheostosis, metaphyseal dysplasia and osteopathia striata. Pycnodysostosis can be differentiated from osteopetrosis and other differentials by its typical appearance of skull, mandible and hands (13). Osteopetrosis is characterized by involvement of medullary canal which is spared in pycnodysostosis. Though hypoplastic clavicles can also be seen in cleidocranial dysplasia, the latter is characterized by osteopenia and osteoporosis. The classical imaging findings of the differentials are summarized.

CONCLUSION

Pycnodysostosis though rare is often diagnosed clinically and with the help of classical imaging findings. Unlike other sclerosing dysplasias patients with pycnodysostosis usually lead a normal life span, however hyperplasia of the uvula may rarely lead to hypoventilation and sudden cardiac arrest.

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DIFFERENTIAL DIAGNOSIS	DIFFERENTIATING FEATURES
1. Osteopetrosis	Medullary canal is affected Anaemia and Splenomegaly are present. Bone in bone appearance No delayed closure of cranial sutures No phalangeal and clavicle hypoplasia.
2. Cleidocranial dysplasia	Predominantly affects intramembranous ossification. Osteopenia & Osteoporosis are often seen, not osteosclerosis.
	 Widened sagittal sutures and/or fontanelles.
	 Premature fusion of the coronal suture.
	 Hypoplasia/aplasia of the lateral clavicle (absent clavicles)
3. <u>Melorheostosis</u>	 Periosteal cortical thickening is characteristic.
	 Thick undulating ridges of bone, reminiscent of molten wax (dripping wax appearance or flowing candle wax appearance)
Metaphyseal dysplasia (Pyle's disease)	 Characterized by flaring of the ends of long bones with relative constriction and sclerosis of the diaphysis and mild cranial sclerosis.
5. Osteopathia striata (Voorhoeve disease)	 Radiographically prominent vertical striations predominate in the metaphyses and epiphyses of the long bones (celery stalk metaphysis).

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OUR EXPERIENCE OF PREGNANCY MANAGEMENT IN A WOMAN WITH KARTAGENERS SYNDROME



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INTRODUCTION

Kartagener's syndrome is an autosomal recessive (AR), ciliary motility disorder characterised by the triad of situs inversus, chronic sinusitis, and bronchiectasis. This rare disorder produces defective movement of cilia which leads to recurrent chest infections, ear, nose, throat symptoms, and infertility. Since the disorder itself is rarely associated with subnormal fertility and infertility the obstetric evaluation and management during pregnancy, labour care and postnatal care is challenging both for the patient and treating Doctors.

CASE SCENARIO

MRS A, 44-year-old with a known case of Kartagener syndrome with infertility, came to our hospital for pre conceptional counselling regarding the risk of IVF and pregnancy management following IVF.

We obtained cardiologist opinion and pulmonologist opinion regarding fitness for IVF and further pregnancy management. Her elder brother was also affected with a similar disease with infertility. She was diagnosed with a case of KARTAGENER'S SYNDROME at the age of 10, when she had multiple episodes of cough, cold. ECHO Findings- SITUS INVERSUS, DEXTROCARDIA, EJECTION FRACTION 65%, NORMAL RV

FUNCTION NORMAL LV FUNCTION. Her pulmonary function test was done from an outside consultant, he has given fitness for IVF and pregnancy management. She was diagnosed to have bronchiectasis – she was on a steroid (fortacort) inhaler. She underwent IVF treatment and had twin pregnancy and underwent prophylactic contical engiselage after NT scan and

She underwent IVF treatment and had twin pregnancy and underwent prophylactic cervical encirclage after NT scan and referred to us for further pregnancy management. They took a rental home near our hospital and came for follow up

In addition to Kartagener syndrome, she had hypothyroidism on oral thyroxine, T2DM on Metformin 500mg bd, developed PIH and started on T. Labetolol 100mg od initially, later required bid dose.Patient was on Fortacart inhaler for cough, Eltroxin 100 od, Metformin 500mg bd, Aspirin 150mg od and Progesterone (C.Susten) 200 bd (oral) Inj Heparin 40 units daily, Inj. Maintane 250 mg weekly Around 24 weeks of gestation one twin had reduced diastolic flow and had IUGR. This twin became IUD at 26 weeks of gestation. Another twin doppler was normal. They were DCDA twins. Around 29 weeks of gestation pt developed severe preeclampsia (urine albumin 3+) with pedal oedema grade-4 pedal oedema (rebound between 2 to 3 minutes with an 8 mm pit). Doppler showed REDUCED DIASTOLIC FLOW for the alive twin. In view of severe preeclampsia, with imminent symptoms, MqSO4 started and steroid prophylaxis was given (we were able to give only one dose) and was taken up for emergency LSCS.

12 hours before admission she took her LMWH. Epidural was not given, risk of spinal haematoma explained during SPINAL ANESTHESIA. Risk of difficulty in extubating and need for ventilatory support explained if general anaesthesia was given.

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The Patient's husband opted for spinal anaesthesia.

Pt had some unknown anti - red cell antibody, so no blood was compatible with her blood during cross matching. Around 25 blood samples were tried. Nothing matched with her blood. We counselled the patient and her husband that if she has pph, we cannot give inj. Prostodin, blood also not available so we may need hysterectomy to save the life of the patient. Mother was safe following surgery, and the baby admitted to the NICU. Alive female baby weight - 870 grams. 1 min Apgar 7/10. Under ventilatory support for 1 day and was given 1 dose of surfactant and was extubated next day.

After the LSCS patient was shifted to ICU, supplemented with oxygen, chest physiotherapy initiated, nebulization given. Early mobilization done, oral feeds started, fluids restricted. 2nd POD shifted to normal ward. Thromboprophylaxis given. Antihypertensives continued. Discharged on 5th POD. Baby was discharged after 2 months with a weight of 1.415kg.

DISCUSSION

Sleigh et al.1, had shown that the cilia are not absolutely immotile, but they have abnormal movements and the secretions of cells of glands could not be effectively expelled, consequently inducing bronchiectasis. Hence this genetic disorder was termed as Primary ciliary dyskinesia (PCD) or dyskinesia cilia syndrome (DSC). KS is a subtype of PCD, referring to PCD cases accompanied with visceral ectopia.

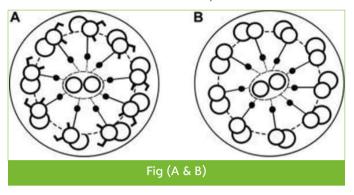
KS tends to run in families, and could occur in the same or different generations, and the parents of most patients are close relatives. The incidence of PCD is about 1/20,000; meanwhile, studies have reported an incidence of birth defects patients with PCD is about 1/20,000-1/60,000.

The first case of bronchiectasis with visceral inversion was reported by Siewart2 in 1904. Four cases with the triad of complete visceral inversion, bronchiectasis, and paranasal sinusitis were identified by Kartagener in 1933. Extensive clinical assessments, with next-generation DNA sequencing and bioinformatics analysis is needed to identify pathogenic genes.

As we all know, cilia are abundant in the respiratory tract, middle ear, oviduct, spermatic flagellum, and brain and spinal ependyma, KS can present with pneumonia, conductive deafness, ectopic pregnancy, infertility, and hydrocephalus.

Other names of Kartagener syndrome (KS) are visceral inversion-naso sinusitis-bronchiectasis syndrome or familial bronchiectasis. KS onset mainly occurs in childhood, but this disease is often misdiagnosed. The major clinical presentations of KS are repeated cough, expectoration, haemoptysis, nasal congestion, running nose, dizziness, and headache. In

addition, some patients may seek healthcare services for infertility. Imaging could provide important evidence for the diagnosis of KS. Indeed, chest X-ray, CT scanning, and ultrasound examinations could identify visceral inversion.



Normal cilia (A) compared with cilia in Kartagener syndrome with missing dynein arms (B).

Male patients are typically infertile which leads to totally immotile spermatozoa. Whereas in females, fertility is more variable, as both fertile and infertile patients were reported.

Women with PCD3 are at risk of infertility and ectopic pregnancy presumably due to defective ciliary function in the fallopian tube.

The present authors searched all published literature and references from relevant articles. Only six cases in four relevant articles were found.

Once a woman has successfully conceived, PCD is unlikely to adversely affect the course and outcome of pregnancy.

However, with the diaphragm pressed upward due to the enlargement of the uterus during the gestational course, the underlying respiratory symptoms will become worse.

The patient may suffer from aggregated and increased frequency of episodes of dyspnoea and cough during aestation, which may increase obstetrical risk during delivery

PREGNANCY MANAGEMENT IN PATIENTS WITH KARTAGENER'S SYNDROME

It is a multidisciplinary approach requiring team of pulmonologist, cardiologist, intensivist, neonatologist

The physiological reduction in lung volume and increase in oxygen consumption during pregnancy may impair respiratory function. The pulmonary function before pregnancy decides the morbidity and outcome during pregnancy and following delivery. The mode of delivery and anaesthesia technique should be planned with a multidisciplinary team. Combined epidural and spinal anaesthesia can be used as it produces pain relief, optimizing respiratory function. Positioning the patient in the OT table - slight reverse Trendelenburg position.

Postoperative period ICU care needed, supplemental

oxygen, chest physiotherapy, postural drainage of bronchopulmonary secretions from the tracheobronchial tree. Nebulization. A good neonatal team is needed for newborn care.

CONCLUSION

Obstetric care for uncomplicated pregnancy itself is challenging. Giving care for women with rare diseases like KS with pregnancy is even more complicated and demanding. Anything can go wrong at any point of time. Advanced planning, detailed communication, and discussion to individuals and their family and involvement of specialists as a multidisciplinary team will improve obstetric outcome, reduce the complication, and save the mother and baby.

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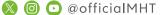
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புற்றுநோய்க்கு எதிரான போராட்டத்தில் நாம் எடுத்து வைக்கும் ஒவ்வொரு அடியும் வெற்றிதான்! நம்பிக்கையுடன் இருப்போம், புற்றுநோயை வெல்வோம்.

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

சிறப்பம்சங்கள்

- அனுபவம் வாய்ந்த கதிரியக்க சிகிச்சை நிபுணர், புற்றுநோயியல் நிபுணர், புற்றுநோய் அறுவை சிகிச்சை நிபுணர் மற்றும் நோய் கண்டறியும் நிபுணர், ஆகியோரை உள்ளடக்கிய புற்றுநோய்க்கான சிகிச்சை குழு.
- மேம்பட்ட வெளி கதிர்வீச்சு நுட்பங்கள் (IMRT, IGRT, ELECTRON THERAPY) மூலம், குறைந்த பக்க விளைவுகளுடன் **35000** க்கும் மேற்பட்ட சிகிச்சைகள்.
- சென்னை போன்ற பெரு நகரங்களில் மட்டுமே இருந்து வந்த பிரேக்கி தெரபி கதிர்வீச்சு சிகிச்சை, தஞ்சை மீனாட்சி மருத்துவமனையில் 2016 ஆம் ஆண்டு அறிமுகப்படுத்தப்பட்டு 2000 க்கும் மேற்பட்ட நோயாளிகள் பயன்பெற்றுள்ளனர் என்பது குறிப்பிடத்தக்கது.





- துல்லியமான மருந்துகளுடன், புற்றுநோயால் பாதிக்கப்பட்ட ஒவ்வொரு நபருக்கும் தனிப்பட்ட சிகிச்சை திட்டங்கள்.
- பகல்நோ பராமரிப்பு பிரிவு (DAY CARE) மூலம் 15000 க்கும் மேற்பட்ட நோயாளிகளுக்கு குறைந்த நேரத்தில் கீமோதெரபி, கதிர்விச்சு மற்றும் வலி நிவாரண சிகிச்சைகள்.
- புற்றுநோய் சிகிச்சைக்கு வயது ஒரு தடையல்ல. ஏனெனில் 80 மேற்பட்ட நோயாளிகளும் குறைந்தபட்ச பக்கவிளைவுகளுடன் எங்கள் மருத்துவமனையில் புற்றுநோய் சிகிச்சை பெற்று குணமடைந்துள்ளனர்.

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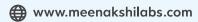
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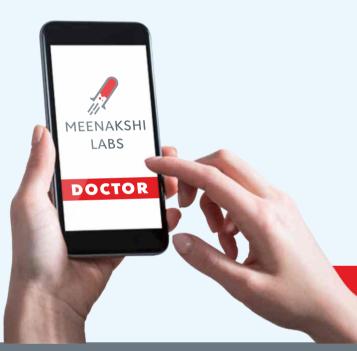


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