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2. David C. Preston MD FAAN, Barbara E. Shapiro. Electromyography and Neuromuscular Disorders: Clinical-Electro physiologic Correlations. ISBN-13: 978-1455726721; Edition: 3: December 5, 2012.
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Editorial

The International Rehabilitation Forum: Growing New Leadership for Rising Countries

One billion people. The population of India. Also the number of people on this planet who suffer from significant physical disability. (1) The readers of the Indian Journal of Physical Medicine and Rehabilitation understand that medical rehabilitation is proven to have a huge impact on the abilities and quality of life of these people. However, since most of these people live in low-resource countries, and since the vast majority of persons in low-resource countries receive no rehabilitation whatsoever, their full potential is not reached. The personal tragedy is seen daily in our clinics. However the burden of 15% of populations who have untreated disability impedes whole societies from reaching a level of comfortable living. Around the globe, as experts in rehabilitation, we have an obligation and an opportunity to change policy and practice. How?

Strategies include our daily practice, research, and mentoring. Also advocacy within our countries. Certainly global organizations like the World Health Organization and the International Society for Physical and Rehabilitation Medicine provide opportunity to reach across the world. However industrialized countries have disproportionate influence on everything from global policy to the research published in medical journals. So there is a very strong need to focus separately on strategies that work for low-resource regions.

The International Rehabilitation Forum (IRF, www.rehabforum.org) was founded exactly to address that need. Its beginnings stem from India. Founded by Indian/American Physiatrist Meeta Peer M.D., and her husband Devendra Peer, CPA, my brother Tom Haig (a well-known disability advocate and broadcast journalist who has lived in India) and Ms. Sierra Loar, MPA, along with myself, the IRF is a very unusual organization. There are no dues (it is free!) and no obligations. Currently no sponsors, either, so it has no money! Legally it is not a membership organization, but rather a registered charity in the United States. People 'join' through the website or 'friend' the IRF through its facebook page. What kind of organization is that? A very, very effective one. That is because we have a singular focus and a creative, collaborative approach.

Examples? When we found that there were only 6 PM&R physicians in all of Sub-Saharan Africa, we made a joke of it: 'The White Book on Physical Medicine and Rehabilitation in Africa and Antarctica'. (2). Except the 'joke' was published in 5 journals around the world as a sign of global alarm and has been cited repeatedly in World Health Organization and other policy documents, as a call to strengthen PM&R throughout the world. When we found that some trainees in Pakistan rescued a few hundred spinal cord injured persons after an earthquake, we encouraged them to publish their work, and called the first world congress on disaster rehabilitation. (3) The first author of that paper is now on our board. We've focused on building rehabilitation in Africa. (4) Not by donating funds, but by understanding the needs and the economics, then encouraging leaders into realizing the need; training rehabilitation scientists, and finding long term mentors. So this year the Ghanaian president announced that his government will build a national rehabilitation center. He didn't do this because a church or foreign government wanted it or because someone donated a facility. But because he knows that his people have a need that is not met. This is sustainable rehabilitation.

One of the IRF's great challenges is South Asia. The region is not without expertise, since some world's great figures in rehabilitation come from South Asia. However the vast majority of people with disabilities here still do not receive PM&R care. The reasons are complex—political, financial, and educational. To begin to address the complexities, we held our Second World Congress in Dhaka, Bangladesh in December of 2012. The strongest leaders of the region, including this journal's editor, met to find a way forward together. Subsequent meetings have led to development of strategies to build the specialty in this region: A consortium of South Asian leaders. A White Book outlining the scope of the field here. A group studying the ways India can build its cadre of PM&R specialists

exponentially to meet the need. We have focused research and educational efforts by many from India and many who care deeply about India. The process is working here! PM&R is strengthening.

The strength of the IRF is not its leadership, but the creativity and drive of its members. The organization is a useful tool for the experts who need to meet together, to share clinical techniques for low-resource regions, and to present their ideas as a unified international organization. Work gets done through the IRF. So we encourage readers who want change to 'join' the IRF. Share your research at our meetings. Get active on our Facebook, with stories, challenges, and ideas for each other. Work with us and your regional leaders on improving the big picture.

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The new Editorial Board has started functioing from 1st April 2013.

Editor along with few members attended 7th World Congress of ISPRM at Beijing, China as faculty and delegates from India, a member country of ISPRM. Exchange of views and updated scientific knowledges were the main event of the conference. We are glad to inform you that lots of renowned physiatrists came from all over the world, appreciated the IJPMR and took keen interest for the journal. They also happily agreed to be an International advisors for the IJPMR. Prof. Andrew Haig is the first one to come forward with his ideas in this issue, though others are not left behind. Editorial Board is thankful to all the members of National and International advisors.

Prof. RN Halder
Editor
IJPMR

Case Report

A Case Report of Hirayama Disease

Kumar Deepak[1], *Gogia Virinder Singh*[2]

Abstract

Hirayama disease is also known as juvenile muscular amyotrophy of distal upper limb. This type of condition is rare and difficult to differentiate this disease from other disease with similar symptoms such as motor neuron disease. Very few cases reported in Medical science and its management is not much proved. But in our case a 20-year-old male came to department with complaint of slowly progressive muscle atrophy and weakness in distal U/L of right side. This disease had confirmed by MRI of flexed cervical region. Patient was managed by similar protocol: Vitamin B12, exercise of hand, cervical collar and isometric neck muscle exercise. Got benefit to patient for cessation of progressive of disease. It is necessary to early diagnosis of Hirayama disease so that patient got early management. Surgical management is preserved to late stage. In case of similar symptoms of cold paresis, amyotrophy, weakness of distal upper limb then a flexion MRI study should be performed to confirm the diagnosis.

Keywords: Hirayama disease, Monomelic amytrophy, Juvenile muscular amyotrophy.

Introduction:

Hirayama disease is a rare disease predominantly affecting the anterior horn cells of the cervical cord in young persons. It is cervical myelopathy characterised by insidious onset of unilateral distal dominant upper limb muscle weakness and atrophy due to anterior cervical cord compression. It has similar symptoms such as motor neuron disease and it is confirmed by magnetic resonance imaging (MRI) in flexed position. Hiryama disease has been reported mainly from Asia and in small number from the western countries[1,2]. This disease, also called as juvenile muscular atrophy of distal upper limbs[1,3], brachial monomelic amytrophy, benign focal amytrophy, is a focal motor disease affecting mainly young people in teen age/early twenties[4]. There is characteristic presentation of weakness and atrophy in one or both upper limbs, followed by spontaneous arrest within several years[5]. Early diagnosis is important because early application of hard cervical collar to minimise neck flexion may prevent disease progression[6].

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Case Report:

This is a case of Hirayama disease in 20 years old male presenting with complaint of weakness of right hand and forearm for more than one and half years on first visit. The weakness was of progressive in nature with muscle atrophy. Recently he had noticed aggravated weakness of right hand and forearm along with tingling and fasciculation in right ring and little fingers. There was no definite history of trauma except few uneventful childhood traumas which were not contributory to present condition. Weakness in grip and pain on medial side of elbow used to increase on exposure to cold (cold paresis). He did not have any past or concurrent history of any other systemic disease. His family history was also not contributory. Patient did not have addiction to any drugs, tobacco or alcohol. Growth history was within normal limit. His hobby was playing cricket and watching movies on TV. A psychological assessment and counselling of patient and his father was also done to address the compliance to rehabilitation management issues.

Neurological examination showed significant atrophy of

muscles on his right forearm and hand including thenar, hypothenar and introsseous muscles (Figs 1 & 2). There were fine irregular and asynchronous tremors in right fingers while extending the fingers (contractile fasciculation). Patient had also complaint of pain and tingling sensation in right little, ring and middle fingers associated with particular type of movement of neck. Manual muscles test of right upper limb showed weakness as compared to left upper limb (Table 1). Deep tendon reflexes of right upper limb were diminished. There was no ataxia, extra pyramidal signs, abnormalities in sweating and urination. Sensations (light touch, deep pressure, vibration, proprioception and pin-prick) were intact.

Laboratory investigation such as CBC, ESR, electrolyte serum Ca^{++}, PO_4^{++}, vitamin D_3 and alkaline phosphatase was within normal limit.

In plain x-ray no any significant abnormalities seen except loss of lordotic curve. MRI cervical spine showed on flexion anterior imaging anterior shift of the posterior dural sac which was noted in the C6-C7 region causing mild flattening of the cord with enhancing posterior epidural mass seen on the contrast image (Figs 3 & 4) which appears to be extending from C4 level till the dorsal spine (Fig 4). Thecal sac diameter at intervertebral disc levels is shown in C4-C5 and C6-C7 (Table 2).

There is EMG and NCS done after clinical examination. Median nerve compound muscle action potential (CMAPs) was reduced and distal latency, F wave latency within normal range. Conduction velocities in all nerve were within normal range (Table 3). Sensory NCS was normal in all nerves (Table 4). In EMG flexor digitorum indices shows active denervetion in the form of there is fasciculation and fibrillation present and chronic nerve denervetion present in the form of neurogenic changes in C7, C8, T1 in myotomes. Extensor digitorum indicis showed no spontaneous activity, there were large polyphasic motor units with reduced recruitment. EMG of C5, C6 myotomes, namely deltoid, biceps brachi and brachiradilalis was normal. This features were suggestive of Hirayama disease.

Discussion:

Hiryama disease mostly found in young adult male and it involves frequently upper extrimities[7,8]. Hypothesized aetiology is as disproportionate growth of vertebral column and content of spinal canal during his growth period. Disproportionate growth affects anterior cervical cord compression due to flexion of neck[9,10]. The mean age is after 15 to 20 years. The onset age is approximately 2 year later than the peak age of the normal growth curve. In our cases onset age is 18 years which is 1 year later than his peak growth.

There are characteristic features of Hiryama disease ie, weakness and atrophy seen in distal and dominant hand muscle (brachiloradialis muscle is relatively spared)[11], young age of onset (10 to teen age of years), unilaterally dominant hand symptoms (rarely B/L hand involvement), onset be stationary after slow progression in years, sensory involvement absent and no lower extremities involvement, no other disease like sringomyelia , spinal cord tumours, MND etc. In this case we found that asymmetry is one of the most characteristic finding of this disease. So adolescent onset of distal upper limb weakness, the finding of asymmetric cord atrophy of flexion MRI studies confirm the diagnosis[11]. The

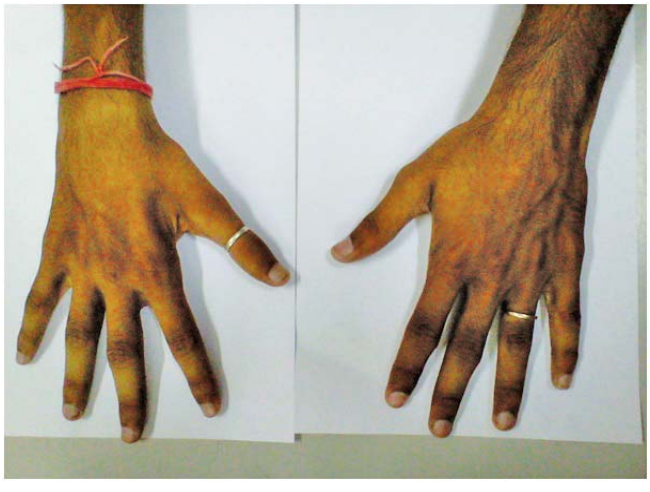

Fig 1- *Showing Significant Atrophy of Forearm Muscles*

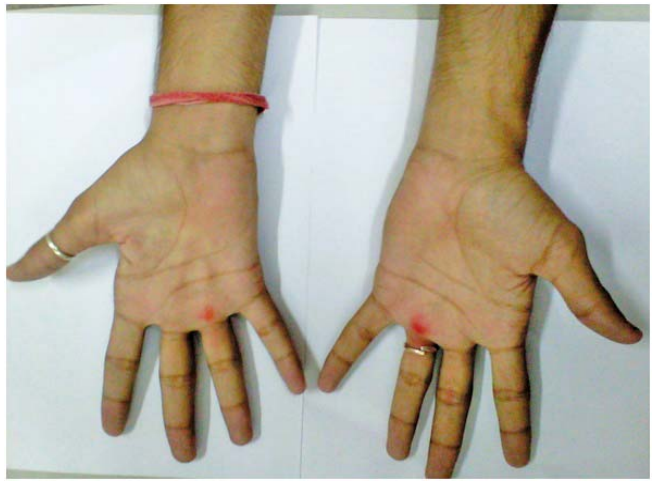

Fig 2- *Showing Atrophy of Thenar Muscles*

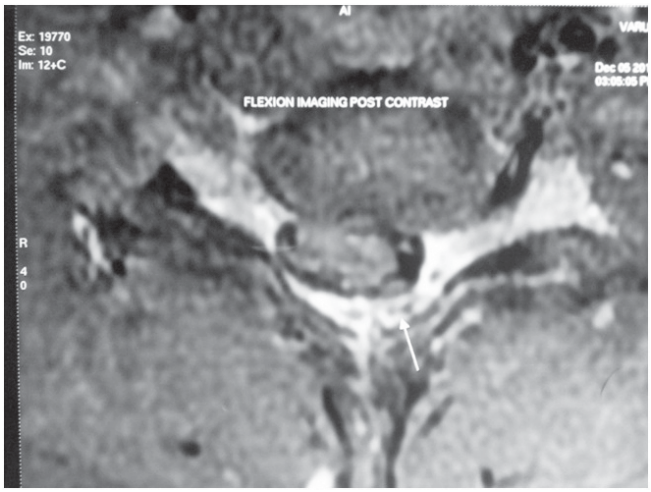

Fig 3- *Showing Posterior Epidural Mass in MRI*

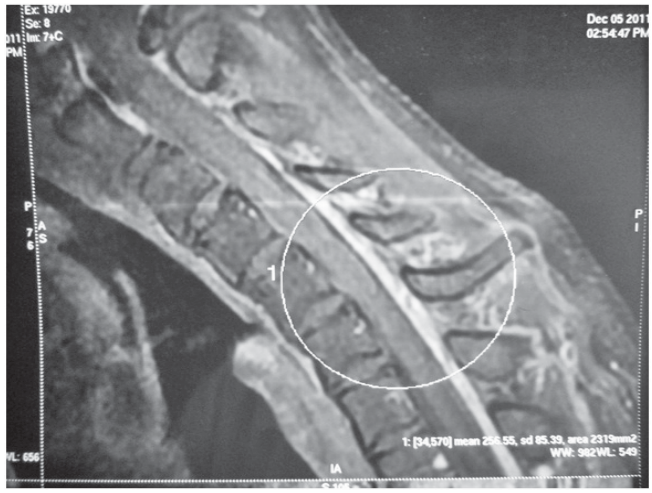

Fig 4- *Showing the Mass in MRI*

Table 1: *Manual Muscle Test*

Movement	Pre-treatment	Post-treatment (follow-up)
Elbow flexion	R4+; L5	R4+; L5
Elbow extension	R 4+; L5	R4+; L5
Wrist extension	R4+; L5	R4+; L5
Wrist flexion	R3+; L5	R3; L5
Supination	R4; L5	R4; L5
Pronation	R4; L5	R4; L5
Hand grip	R Poor; L Good	R Poor; L Good
Finger abduction	R3+; L5	R3+; L5
Finger adduction	R3+; L5	R3+; L5
Forearm girth	R 23cm L 26 cm	R 24cm; L 28 cm
Cold paresis	R Present; L Absent	R Present; L Absent
Contractile fasciculation	R Present; L Absent	R Present; L Absent
Static tremor	R Present; L Absent	R Present; L Absent

R: Right side; L: Left side; NA: Not available

pathophysiology of repeat cervical cord trauma (atrophy) due to neck flexion allow to prevent flexion of neck by hard cervical collar therapy. Early hard cervical collar therapy arrest the induces a premature arrest shorten duration of illness, also helpful to minimise functional disability[11]. Few cases which is not responsive to conservative treatment undergone to surgical treatment anterior cervical decompression and fusion got benefited[11]. For advance Hirayama disease, tendon transfer improves the activities of daily living[12]. In our case patient was advised for hard cervical collar therapy, vitamin B12 supplement along with strengthening exercise of right U/L and isometric neck muscle exercise. Patient has responded well and no further muscle weakness, cold paresis and fasciculation worsening seen.

Conclusion:

We should keep in our mind in case of insidious onset of unilateral distal upper limb weakness in young man with cold paresis and contractile fasciculation, the finding of asymmetric lower cervical cord atrophy on routine MRI studies suggestive of Hirayama disease. There should be cervical flexion MRI study performed to confirm the diagnosis. Isometric neck muscle exercise is not proving for beneficial and requires more work over there. There should be early diagnosis because early use of hard cervical collar application to reduce neck flexion has

Table 2: *Thecal SAC Diameter at Intervetebral Disc Levels*

Intervetebral disc level	Antero-posterior diameter
C2-C3	12.0 mm
C3-C4	11.0 mm
C4-C5	10.0 mm
C5-C6	11.0 mm
C6-C7	10.0 mm

Table 3: *NCV (Nerve Conduction Velocity)*

Nerve and site	Latency	Amplitude	Segment	Latency difference	Distance	Conduction velocity
Median	**Nerve**	**Right**				
Wrist	4.0ms	5.7mV		ms	mm	m/s
Elbow	7.4ms	5.5mV	Wrist-Elbow	3.4ms	200mm	59m/s
Axilla	10.5ms	5.0mV	Elbow-Axilla	3.1ms	210mm	68m/s
Erb,s point	14.0ms	4.9mV	Axilla-Erb,s point	3.5ms	200mm	57m/s
Ulnar	**Nerve**	**Right**				
Wrist	3.3ms	4.7mV				
Below elbow	6.4ms	4.9mV	Wrist-Below elbow	3.1ms	170mm	55m/s
Above elbow	8.9ms	4.7mV	Below elbow-above elbow	2.5ms	110mm	44m/s
Erb,s point	14.7ms	3.1mV	Axilla-Erb,s point	3.5ms	180mm	51m/s
Median	**Nerve**	**Left**				
Wrist	3.7ms	9.5 mV				
Elbow	6.7ms	9.5mV	Wrist –Elbow	3.0ms	200mm	67m/s
Axilla	9.9ms	8.6mV	Elbow – Axilla	3.2ms	210mm	66m/s
Erb,s point	13.0ms	7.9mV	Axilla - Erb,s point	3.1ms	200mm	65m/s
Ulnar	**Nerve**	**Left**				
Wrist	2.9ms	9.1mV				
Below elbow	5.9ms	8.2mV	Wrist - Below elbow	3.0ms	170mm	57m/s
Above elbow	8.1ms	6.4mV	Below elbow - Above elbow	2.2ms	110mm	50m/s
Axilla	10.8ms	6.2mV	Above elbow - Axilla	2.7ms	150mm	56m/s
Erb,s point	13.8ms	5.7mV	Axilla – Erb's point	3.0ms	180mm	60m/s

Table 4: *Sensory Nerve Condition*

Nerve and site	Peak latency ms	Amplitude Micro volt	Segment	Latency difference Ms	Distance Mm	Conduction velocity m/s
Median	**Nerve**	**Right**				
Wrist	4.1	33	Index - wrist	2.5	150	60
Ulnar	**Nerve**	**Right**				
Wrist	2.8	24	Little finger - wrist	2.3	230	56
Median	**Nerve**	**Left**				
Wrist	3.4	39	Index - wrist	2.7	150	56
Ulnar	**Nerve**	**Left**				
Wrist	3.9	31	Short finger - wrist	2.5	130	52

been shown to prevent progressive muscular weakness and stop disease progression.

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Case Report

Mucopolysaccharidosis and Rehabilitation

Harshanand P[1], Anil Kumar G[2], Vivek P[3], Jayasree R[4]

Abstract

Mucopolysaccharidosis is a rare lysosomal storage disorder with overall prevalence of all types is 3.53 per 100,000 live births. Exact figures are not available for Indian population. It has poor prognosis with no easy curative medical or surgical management. This case report describes two cases of mucopolysaccharidosis with type one and type four variant. These cases diagnosed and rehabilitated to increase quality of life. Early identification of such cases by clinical features, supportive investigations and rehabilitation management can help patient to improve functional independence and activities of daily living.

Key words: Mucopolysaccharidosis, Rehabilitation.

Introduction:

The mucopolysaccharidoses (MPSs) are a family of metabolic disorders caused by the deficiency of lysosomal enzymes needed to degrade glycosaminoglycan (GAG)[1]. GAG is an important constituent of the extracellular matrix, joint fluid, and connective tissue throughout the body. Progressive accumulation of GAG within the cells of various organs ultimately compromises their function. The overall prevalence of all types of MPSs is 5.53 per 1,00,000 live births[2].

Case 1:

A 10 year old male child came to PMR OPD with chief complaints of dysmorphic facial features since birth and regression of milestones. It was associated with decreased vision and intelligence. There was history of consanguineous marriage (first degree). He was operated 2 years back for umbilical hernia. On clinical examination, more abdominal girth, mild hepato-splenomegaly, and mild cardiomegaly were present. CNS examination: Higher function- speech and intelligence is moderately impaired with long term memory loss. Cranial nerve examination, vision is impaired due to corneal clouding. Motor examination showed power of all muscles was 3/5. Reflexes of upper and lower extremities were brisk with Babinski reflex positive. Musculoskeletal examination showed characteristic facial appearance (Fig 1) which includes macrocephaly, bossing of forehead, proptosis of both eyes, corneal clouding, low set ears, flattening of nose bridge, macroglossia, widening of teeth, short neck, short chest, kyphoscoliotic deformity with convex curve towards right side, widening of metaphysis at elbow, wrist, knee and ankle seen. Both hands showed short phalanges. Range of motion at shoulder, hip restricted bilaterally. Elbow and knee range of motion showed fixed flexion deformity of 20 degree. On functional evaluation child was having good sitting balance, he was unable to do all routine activities of daily living like eating brushing, grooming, clothing and toiletting because of deformity. On gait analysis; he walks with support to both hands, hip and knee in flexion, ankle with foot in dorsiflexion. Patient was investigated with radiographs. Radiographs of skull (Fig 2) showed anteroposterior diameter more than lateral, 'J' shaped sella tursica, premature fusion of sutures, and widened mandibular angle, dorsolumbar spine (Fig 3) showed kyphosis, lumbar vertebrae showed anteroinferior beaking with posterior scalloping. Ribs

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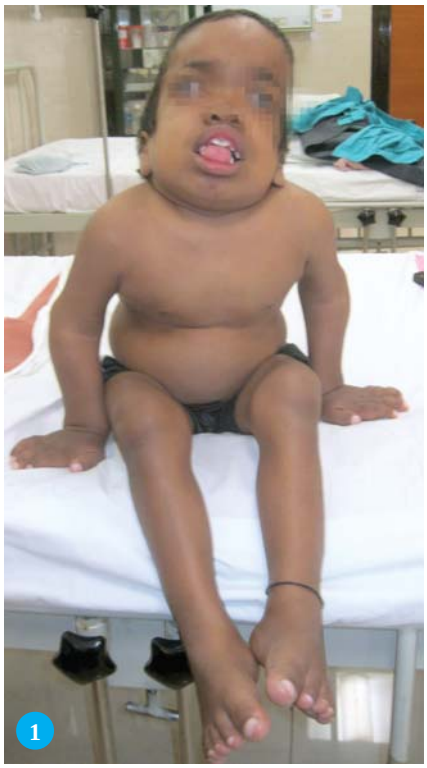
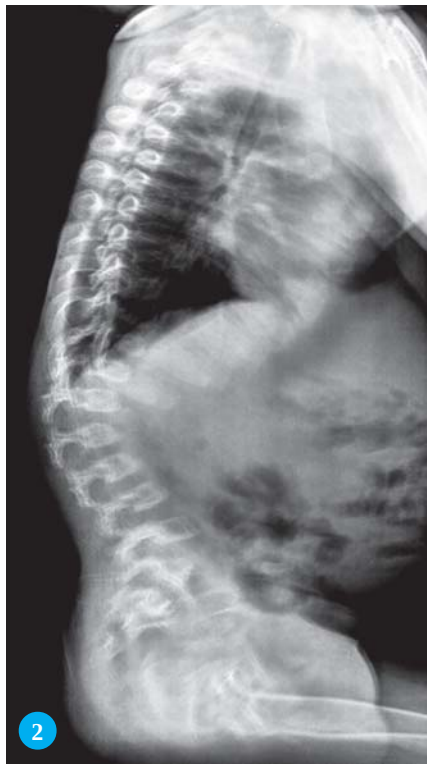
were wide and paddle shaped. Radiographs of elbow and hand showed widened diaphysis of long bones with osteoporosis and cortical thinning, subluxation of proximal radioulnar joints, proximal tapering of metacarpals with short and broad phalanges. Radiographs of knee showed bilateral medial bowing of lower end of femur. Urine analysis for screening of mucoplysaccharidosis was positive. Blood examination for typing/ deficiency of enzyme of mucopolyscaccharidosis done and it is positive for alpha-L-iduronidase. By clinical examination and biochemistry, diagnosis of mucopolysaccharidosis type 1 (Hurler syndrome) confirmed.

Case 2:

A 8-year-old female child came to PMR OPD with chief complaint of inability to walk with prominences of bones of upper and lower extremity. She started difficulty in walking with prominence of wrist, elbow and knee bones from 2nd year. She developed weakness of neck and back muscles producing neck hyperextension and kyphoscoloitic deformity at back. Symptoms were progressing slowly. She lost walking and routine activities of daily living like eating, brushing, grooming etc, since last one year. On clinical examination, cardiovascular system showed mild cardiomegaly. Central nervous system: Higher functions were normal. Power was generalised 3/5. Musculoskeletal examination: Facial features included macrocephaly, bluish tinge of sclera, high arched palate, short neck with poor stability or control, (Fig 4) prominent chest ribs, prominent elbow, wrist joints, knee in genu valgum deformity, prominence of knee joint bones. Joints were hypermobile with exceess range of motion at wrist extension, hip extension, internal and external rotation, and ankle joint dorsiflexion.

Radiographs of skull showed similar findings with case one. Radiograph of cervical spine showed odontoid hypolasia. Radiograph of thoracolumbar spine (Fig 5) showed kyphoscoliosis with anteriot beaking of lumbar vertebrae. Radiograph of radius and ulna showed similar findings with case one. Screening of urine for mucoplysaccharides ie. glycos-amino-glycans (GAG) done. Report came positive for mucopolysaccaridosis. Two dimensional electrophoresis of 24 hours urine for typing of mucopolysaccharidosis has been done. Report positive for mucopolysaccharidosis type 3 with advice of clinicopathological correlation. The diagnosis of mucopolysaccharidosis should be confirmed by direct





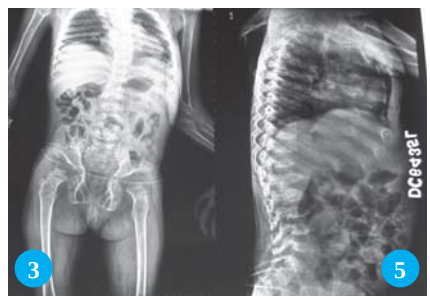


Fig 1- *Showing Characteristic Facial Expression*; **Fig 2-** *Straight X-ray Skull Showing Anteroposterior Diameter more than Lateral*; **Fig 3-** *Straight X-ray Showing Dorsolumbar Kyphosis*; **Fig 4-** *Showing Facial Feature*; **Fig 5-** *Thoracolumbar Spine Showing Kyphoscoliosis*

enzymatic assay in leucocytes or fibroblasts. These tests were not available in India. After clinicopatho-radiological correlation, diagnosis of mucopolysaccharidosis type 4 (Morquio syndrome) has been drawn.

In consideration of availability of medical management (enzyme replacement therapy in india) post surgical complications and life expectancy of bone marrow transplantation[3], rehab management is more significant and helpful to improve quality of life of patient.

Rehabilitation Management:

Counselling of parents about prognosis of the disease done. Genetic screening of parents and other siblings were advised.

Problems list for both cases

1. Generalised weakness of all muscles, dependent ambulation, activities of daily living, transfers and toileting.
2. Poor head control, kyphoscoliotic deformity at back and impaired sitting balance
3. For case one; Impairment of vision due to corneal clouding

Rehabilitation:

Submaximal strengthening exercises of all muscles of body, stretching exercises and deep breathing exercises has been given to prevent contractures and increase vital capacity of lung.

- Night splints have been given to prevent further occurrence of deformity at knee and elbow. Cervical collar have been given for cervical instability and its future complication to both cases. Standard wheelchair with neck support has been given in consideration of future progress of disease. Patient has been trained for activities of daily living with minimal assistance of care giver.
- For case one, as kyphoscoliotic deformity is correctable, posterior shell total contact thoracolumbar orthosis with lateral extension with anterior ring and straps given. Rationale behind giving anterior ring and straps was to prevent chest expansion and respiratory compromise due to orthosis in future. For case two, total contact thoracolumbar orthosis correcting scoliosis with anterior straps has been given.
- For case 2, wrist joint stabilisation of right side done by below elbow cock up splint for finger movement and to increase grip strength. Left side resting hand splint given.
- Patient has been taught transfers from wheelchair to bed/commode and vice versa.
- For case one, Opthalmology consultation done for decreased vision and advised cataract surgery.
- For case one, special school for mentally challenged has been advised. For case two, normal school has been advised.
- The WEE FIM scoring before and after six months of rehabilitation done. It is depicted in Table 1.

Disscussion:

Mucopolysaccharidosis was first described by Charles Hunter, a Canadian physician. Mucopolysaccharidosis type I (MPS I) Hurler syndrome is an autosomal recessive disorder caused by deficiency of a-L-iduronidase. Mucopolysaccharidosis type I have incidences of 0.69 per 100,000 live births[3]. Characteristic clinical features which differentiate it from other types include early onset of disease, dysmorphic facial features, corneal clouding, mental retardation, umbilical or inguinal hernia, and dysostosis multiplexa. All these clinical features were present in case one. Morquio syndrome (Type 4) incidence: Internationally; the estimated incidence covers a wide range, including 1 case per 75,000 births in Northern Ireland, 1 case per 200,000 births in British Columbia, and 1 case per 263,157 births in Germany [4].

Medical management is based on principle of enzyme replacement therapy. Some US based pharmaceutical companies came with new drug known as Aldurazyme® (laronidase)[5]. It is the first enzyme replacement therapy drug to specifically treat the underlying cause of MPS I. It is prescribed for people with Hurler and Hurler-Scheie forms of MPS I and for people with the Scheie form who have moderate to severe symptoms. The main problem with treatment of laronidase is anaphylaxis and related complications. This drug is not available in India. No enzyme replacement therapy available for Morquio syndrome. Vellodi *et al*[3] did study on life expectance post bone marrow transplantation of MPS 1 patients and shows that average duration of life expectancy is 3 years with recurrent bone marrow transplantation.

Very few publications are available about rehabilitation management of mucopolysaccharidosis. Gulati and Agin[6] (1996) studied Morquio syndrome: a rehabilitation perspective. They described rehabilitation of Morquio patients post quadriparesis due to odontoid hypoplasia. Because of odontoid dysplasia, spinal cord compression occurs which leads to tetraplegia or quadriplegia. This

Table 1: *Showing Wee Firm Score before and after Rehabilitation*

Area		Case One		Case Two	
		Before rehabilitation	After rehabilitation	Before rehabilitation	After rehabilitation
Self care	1. Eating	1	6	1	6
	2. Grooming	1	6	2	4
	3. Bathing	1	5	2	4
	4. Dressing– Upper Body	1	5	2	6
	5. Dressing– Lower Body	1	5	2	6
Sphincter control	6. Toileting	7	7	7	7
	7. Bladder management	7	7	7	7
	8. Bowel management	7	7	7	7
Transfers	9. Transfers:Bed/Chair/Wheelchair	1	4	1	4
	10. Transfers: Toilet	3	4	1	4
	11. Transfers: Bath/Shower	3	5	1	4
	12. Walk/Wheelchair	1	6	1	4
	13. Locomotion: Stairs	1	1	1	1
Communication	14. Comprehension	7	7	7	7
	15. Expression	7	7	7	7
Social cognition	16. Social interaction	7	7	7	7
	17. Problem solving	3	3	7	7
	18. Memory	3	3	7	7
WeeFIM® Total		72	96	70	99

article describes rehabilitation of quadriplegic patients of Morquio syndrome.

Conclusion:

The prognosis of Hurler syndrome is poor with average life expectancy for Hurler syndrome is first decade. Death mostly occurs due to respiratory failure. Early diagnosis of patients with appropriate rehabilitation can improve functional independence and activities of daily living.

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Medical Philately

Here is an image of a stamp depicting wheelchair basketball.

Printed by photogravure

Issued by Israel on November 6, 1968 to publicize the International Stoke Mandeville Games.

Scott No. 377.

Charcot Arthropathy in Diabetes

Balamurugesan Kandan[1], Viswanathan Stalin[2]

This 55-year-old lady with type 2 diabetes for the last six years came with complaints of a painless right ankle swelling of two months' duration. There was no prior trauma, fever, or swelling in other joints. Examination revealed a swelling involving the ankle and lower 1/3rd of the leg without warmth or tenderness (Figs 1 and 1). She had a healed trophic ulcer at the base of first toe on the right side, normal peripheral pulses and ankle-brachial index of ~1.0. Vibration and joint position sense was reduced bilaterally.

The association between diabetes and Charcot arthropathy was shown in 1936[1]. Charcot arthropathy is classified into stages 0[inflammatory], 1[development], 2[coalescence] and 3[remodelling][1]. Treatment is generally non-operative and includes off-loading, casts, braces and orthotic support[1,2]. Surgeries like osteotomy and arthrodesis are rarely required. She was referred for physiotherapy but lost to follow up thereafter.

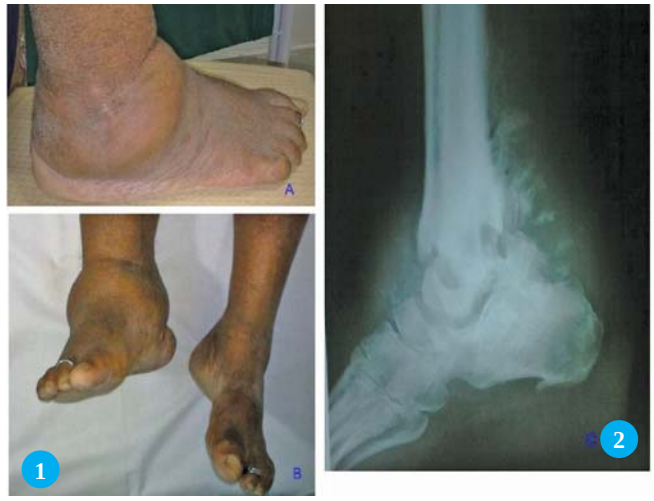

Fig 1- Showing Swollen Right Ankle Joint without Warmth or Tenderness

Fig 2- Ankle Radiography Showing Sclerosis, Destruction of Talus and Tibiotalar Joint

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PG Forum

REHAB CHALLENGES

A 42 years male patient presented with complete flaccid paraplegia following a fall with a D5 neurological level (skeletal level D4) complicated with grade IV sacral infected pressure ulcer (10 x 12 cm), urinary and bowel incontinence along with restricted right wrist ROM due to a fracture. After a prolonged conservative rehabilitation programme along with couple of surgeries like K wire fixation of his right radius and ulna (Fig 1) and pressure sore flap surgery the patient's clinical condition improved significantly. Meanwhile upper limb muscle strengthening, tilt table training and cardiovascular training were carried on. The patient was successful to manage his bladder by CIC himself. But the trunk muscles power was not developed greatly. He is now keen to mobilise himself.

But we noticed that his hips ROM were significantly reduced due to development of heterotrophic ossifications (Fig 2). After six weeks course of postural care, exercise therapy, Risedronate and Indomethacin the HO (Fig 3) were not improved significantly. That's why he has poor sitting posture due to approximate hip ROM of 0- 30 degree flexion (Fig 4).

He is still on conservative regimen for HO but too early for surgical correction. He is not a candidate for KAFO with crutches. Unfortunately he cannot afford a motorised wheelchair for his self mobility.

Please opine regarding best possible mobility devices for him.

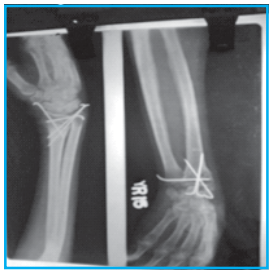

Fig 1

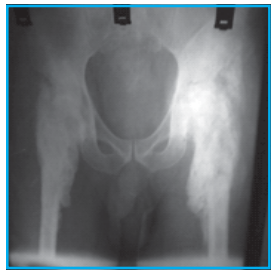

Fig 2

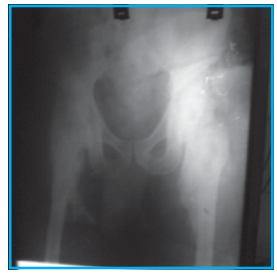

Fig 3

Fig 4

PG Forum

REHAB QUIZ

1. **Which of the following nerves contain no cuteneous sensory fibres?**
   A) Long thoracic
   B) Axillary
   C) Musculocutaneous
   D) Tibial

2. **Fibre density determined by single fibre EMG is increased in all except**
   A) The elderly
   B) Children below the age of 10
   C) Denervation
   D) extensive fibre splitting

3. **The blink reflex is of value in diagnoses of**
   A) Multiple sclerosis
   B) Bell's palsy
   C) Acoustic neuroma
   D) All of the above

4. **A female paraplegic secondary to complete spinal cord lesion can**
   A) Have a vaginal delivery
   B) Experience organic sensation following tactile stimulation of the breast
   C) Have regular menstrual period
   D) All of the above

5. **Which of the following well motivated geriatric patients should not receive prosthesis?**
   A) A/K-B/K bilateral male amputee with good cardiac reserve.
   B) Bilateral A/K amputee with mild hemi paresis who had walked before surgery of the second leg.
   C) Unilateral A/k with vascular disease but no gangrene of the other leg
   D) Unilateral B/K 90 year old female

6. **Tilting of pelvis occurs maximally**
   A) In mid stance
   B) At push off
   C) During swing through
   D) At heel strike

7. **Definite diagnoses of psedogout is made on the basis of**
   A) Presence of mono sodium urate crystal in synovial fluid
   B) X ray evidence of calcification of articular cartilage (chondrocalcinosis)
   C) Presence of calcium pyrophosphate dehydrates crystal in synovial fluid or tissue.
   D) Normal serum uric acid level

8. **Mixed Connective Tissue Disease (MCTD) is characterized by all except**
   A) Reynaud's phenomenon
   B) Positive fluorescent ANA
   C) Normal aldolase level
   D) Oesophageal hypo motility.

9. **Arthrogryposis multiplex congenital characterized by all except**
   A) Decreased muscle bulk
   B) Flexion contracture of wrist
   C) Syndactyly or polydactyly
   D) External rotation of arms

10. **In COPD pulmonary function test will not reveal**
    A) Decreased lung volume and expiratory flow rate
    B) Decreased inspiratory capacity
    C) Increase in maximum breathing capacity
    D) Decrease in carbon mono oxide uptake

ANSWERS

March 2013 issue:

1-C; **2**-A; **3**-B; **4**-B; **5**-B; **6**-D; **7**-C; **8**-D; **9**-B; **10**-B

Case Report

Anatomic Reconstruction of the Distal Radio-ulnar Ligament for Distal Radio-ulnar Joint Instability : A Case Report

Sahoo J

Abstract

Stability of the distal radio-ulnar joint (DRUJ) is provided by bony architecture and by soft tissues such as the triangular fibrocartilage complex (TFCC), the joint capsule, and surrounding muscles. Many authors have made various attempts to restore DRUJ stability surgically following trauma. The objective of this study was to analyse clinical outcomes after anatomic reconstruction of the distal radio-ulnar ligaments in a 26 years male with post-traumatic chronic instability of the DRUJ. Anatomic reconstruction of the major structures responsible for joint stability is the most important principle for the treatment of instability of an injured joint with an intact articular surface by using a palmaris longus tendon graft whose ends were anchored in pre-drilled holes in the radius and the ulna. Anatomical reconstruction of the distal radio-ulnar ligaments is thought to be an effective procedure for treating post-traumatic DRUJ instability.

Keywords: Distal radio-ulnar joint, Triangular fibrocartilage complex, Palmaris longus.

Introduction:

Cadaver studies have demonstrated that the volar and dorsal distal radio-ulnar ligaments, which are components of the TFCC, play a major role in stabilising the radio-ulnar joint[1,2]. Many authors have made various attempts to restore DRUJ stability surgically following trauma. However, these non-anatomic procedures are not reliable[3,4]. Anatomic reconstruction of the major structures responsible for joint stability is the most important principle for the treatment of instability of an injured joint with an intact articular surface. The objective of this study was to analyse clinical outcomes after anatomic reconstruction of the distal radio-ulnar ligaments in a patient with post-traumatic chronic instability of the DRUJ.

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Case Report:

Twenty six years old male had chief complaint of painful motions of the left wrist joint and inability to lift heavy object in that hand. He had history of injury four years back, for which he was treated by crepe bandage application and medication. He had persistent insecurity and discomfort during his daily activities, refractory to conservative management. Then he came to SVNIRTAR on 24.1.11. On examination there is a dimple in the ulnar side on pronation of forearm (Fig 1). The Piano key sign is positive. All the wrist range of movements were within normal limit. No significant tenderness was found.

A 4 to 5 cm curvy-linear skin incision (Fig 2) was made from the distal ulnar head extending proximally along the fifth extensor compartment. The fifth extensor compartment was opened, and the extensor digiti minimi tendon was retracted radially. An L-shaped flap was created in the distal radio-ulnar joint capsule, with one limb made along the dorsal rim of the sigmoid notch and the other made proximal and parallel to the dorsal DRUJ. The palmaris longus tendon graft (Fig 3) was harvested, and each end of the graft was connected to a thread to facilitate easy passage through the tunnels. A 2.0 mm drill hole was made in the radius, and then gradually enlarged with a straight curette to allow free passage of the tendon graft. To make the ulnar tunnel, the wrist was flexed volarly and the TFCC was pulled

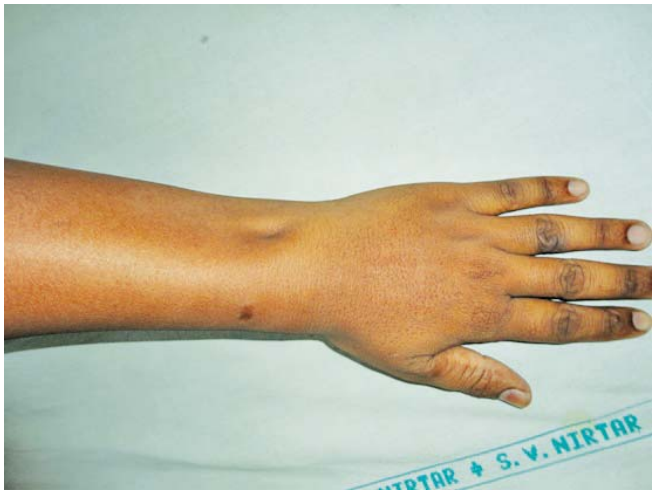

Fig 1- *Showing Dimple*

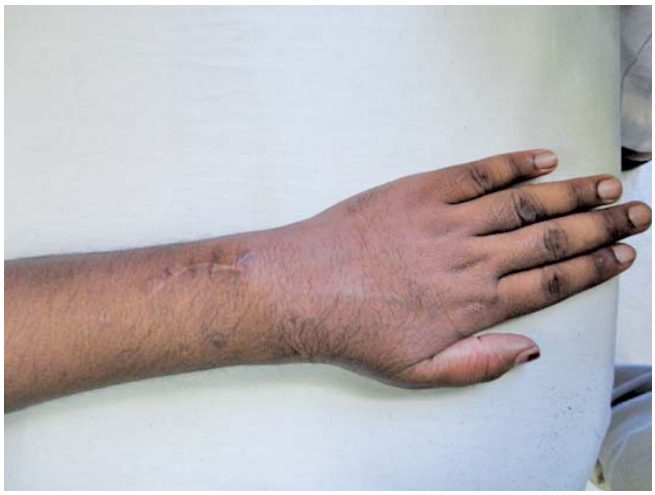

Fig 2- *Showing Skin Incision*

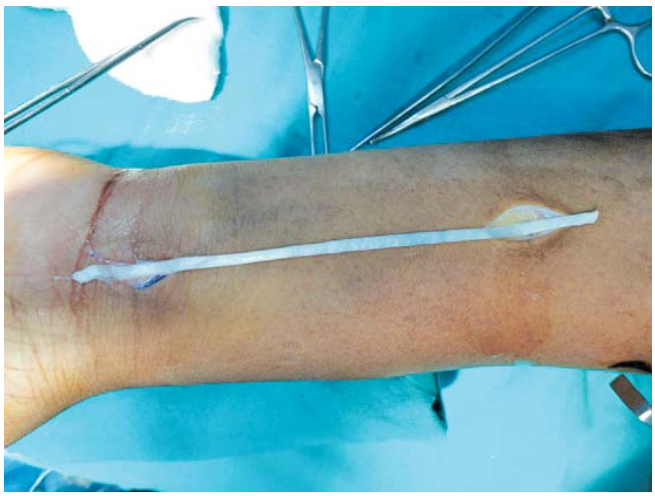

Fig 3- *Showing Graft*

distally to expose the ulnar fovea. With the ulna pulled dorsally, a tunnel was made through the ulnar fovea at a tilt angle towards the ulnar side of the ulnar neck in the same way for the radial tunnel (Fig 4). The tendon graft was passed through the radial tunnel and both of its limbs were brought to the opening in the ulnar fovea. Then both limbs were passed through the ulna tunnel to the ulnar side of the ulnar neck. The limb on the volar side surrounded the ulna and was sutured with the other limb using 3-0 non-absorbable sutures (Fig 5). During the suturing procedure, both limbs were pulled taut with the forearm in neutral position, and care was taken not to restrict forearm rotation by imposing excessive tension. Motion of the wrist joint and the DRUJ was checked, and the final status of the reconstructed tendon was assessed with the anteroposterior stress test. Postoperatively, long-arm cast was applied for 6 weeks, followed by range of motion exercise to restore normal joint mobility. Normal activity was allowed at 10-12 weeks postoperatively, once normal range of motion was achieved.

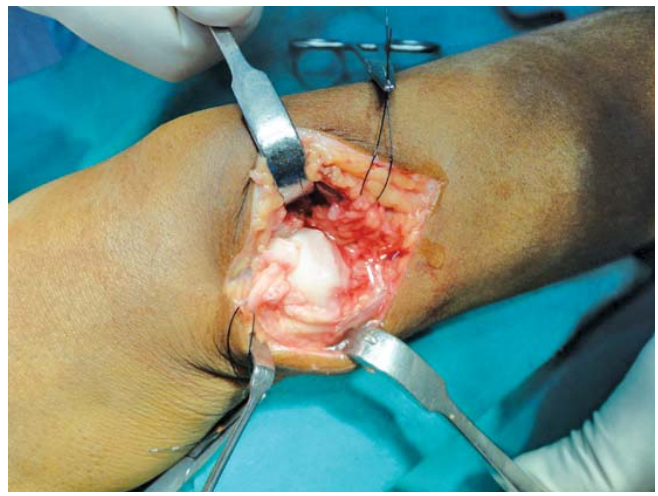

Fig 4- *Showing the Tunnel*

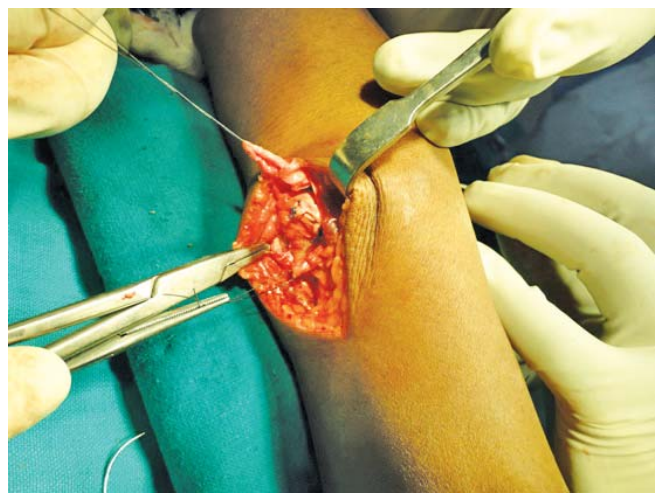

Fig 5- *Showing Suturing*

Objective clinical examination was performed using the anteroposterior stress test, range of motion evaluation, grip strength assessment using a dynamometer and plain radiography. Subjective clinical evaluation was performed using the Patient Rated Wrist Evaluation (PRWE)[5] and the Disabilities of the Arm, Shoulder and Hand (DASH)[6] at six months and one year . Till date patient have no similar complaint as before.

Discussion:

The methods for treating DRUJ instability have evolved from non-anatomical methods such as tenodesis to anatomical methods such as reconstruction of the distal radio ulnar ligaments. As anatomical reconstruction has been proven as the optimal treatment for instability in other joints such as knee and elbow, this attempt is quite natural. The reconstruction procedures suggested by some authors[7,8] using the palmaris longus tendon has been shown to be the most reliable and anatomically acceptable surgical technique. I performed distal radio-ulnar ligament reconstruction accordingly. DRUJ instability is difficult to diagnose because it presents with variable and often indistinct symptoms. The diagnostic criteria of DRUJ instability in the current study included repeated subluxation and reduction or dynamic instability observed during the stress test. The satisfactory results cannot be expected only by the joint realignment in patients with obvious subluxation because the dynamic instability would impair the DRUJ function. It is clear that ligament reconstruction is required when functional disability is largely attributable to dynamic instability. Functional results were satisfactory with preservation of joint motion. At the last follow-up, no objective symptoms or signs of subluxation were noted. Anatomical reconstruction of the distal radio-ulnar ligaments is thought to be an effective procedure for treating posttraumatic DRUJ instability.

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Original Article

Profile of Traumatic Spinal Cord Injury Patients Admitted in Physical Medicine and Rehabilitation Department of a Tertiary Care Hospital: A North East India Experience

Chanu A R[1], Zonunsanga C[2], Hmingthanmawii[3], Pertin M[4], Romi Singh N[5]

Abstract

Study Design: Retrospective Descriptive Study.
Setting: Physical Medicine and Rehabilitation (PMR) Department, Regional Institute of Medical Sciences (RIMS), Imphal, a tertiary care teaching hospital in North East India
Study Duration: 1st November 2011 to 31st October 2012.
Objective: To study the profile of traumatic spinal cord injury (SCI) patients admitted in PMR Department, RIMS.
Materials and Methods: Neurological profile of traumatic SCI patients admitted in PMR Department, RIMS was recorded using a structured proforma and analysed. Demographic profile of the patients, time since injury, functional status and complications were also recorded.
Results: Among all 22 patients, 16(72.7%) were tetraplegics with C5 (59.09%) as the most common neurological level involved. Twelve (54.5%) were American Spinal Injury Association (ASIA) grade A. The mean motor and sensory scores were 45±24.97 and 157.50±69.53 respectively. The mean FIM score (at admission) was 71.50±23.40 and FIM (at discharge) was 82.35±20.72. Spasticity was present in 16 cases (72.7%) with gastrosoleus as most common site. There were 14 patients (63.6%) who had urinary tract infection (UTI). Only 5(22.7%) underwent urodynamic study and all had hyperactive detrussor. The most common mode of bladder management was clean intermittent catheterisation (CIC) which was done in 16 patients (72.7%). Pressure sore was seen in 14(63.6%) of patients with sacrum (78.57%) as the most common site.
Conclusion: Majority of traumatic SCI inpatients were of ASIA grade A and tetraplegics were commoner. Fall from height was the comonest cause of injury. Spasticity, UTI and pressure sores were common complications.

Key words: Traumatic Spinal cord injury, ASIA, Clean intermittent catheterisation, Functional independence measure, Pressure sore.

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Introduction:

Spinal cord injury (SCI) is a major debilitating disease which results in functional, economic, psychological and social disability. The developments in the management of SCI have led to decrease in morbidity and mortality rates, thereby increasing the prevalence of patients with varying degrees of functional limitations. Although patients with SCI have a great impact on healthcare system, there is a dearth of reliable statistics concerning spinal injury in India and more so in this part of the country. There is still a need for more insight into the characteristics of the populations affected with SCI. In this retrospective and descriptive study, we aimed to identify the neurological and demographic profile of patients with traumatic SCI admitted in PMR Department, Regional Institute of Medical Sciences (RIMS), Imphal.

Objective:

To study the profile of traumatic SCI patients admitted in PMR Department, RIMS.

Material and Methods:

Hospital files of traumatic SCI patients admitted for rehabilitation between 1st November 2011 and 31st October 2012 were reviewed retrospectively. Only the data of first admission to our hospital were included in the study. Demographics, aetiological factors, level of injury (ASIA impairment scale), and functional status (functional independence measure: FIM) at admission and discharge were recorded. Complications related with SCI were also noted. Statistical analysis was conducted using SPSS for Windows 17.0. Frequencies, percentages, median and mean values were calculated.

Results:

Demographic and neurological characteristics of patients are presented in Table 1. A total of 22 patients (mean age 40.41±15.18 years, 95.5% male) were included. There was a wide range among the patients in regards to the time of reporting since injury with median time being 34.5(range 9-132) days. Six patients (27.3%) had already undergone some form of surgery viz decompression ± internal fixation before being transferred or admitted to our ward.

Eleven (50%) of the cases were due to fall from height (FFH) and 8(36.4%) were due to road traffic accident (RTA) with the remaining 3(13.6%) resulting from direct hit on the spine as shown in Fig 1. Sixteen (72.7%) of the patients were tetraplegic with C5 (59.09%) as the most commonest neurological level. The mean motor and sensory scores were 45±24.97 and 157.50±69.53 respectively. Twelve cases (54.5%) were of ASIA grade A (complete), 4 (18.2%) comprised ASIA grade C and 6 (27.3%) comprised of ASIA grade D. At the time of admission, cervical motor level (tetraplegic) was the most commonly found neurological level comprising 72.7% in our series as shown in Table 2. Different sensory levels were also shown at Table 2 with 45.5% of the patients having sacral sparing. The mean FIM score at admission and discharge were 71.50±23.40 and 82.35±20.72 respectively . Most of the patients had at least one form of complications during admission as being shown at Table 3. Spasticity was present in 16 cases (72.7%) with gastrosoleus as most common site. There were 14 patients (63.6%) each, who had UTI and pressure ulcer. Other complications observed during admission included orthostatic hypotension (8 patients), 1 case each with shoulder pain, bladder stone and fistula (penoscrotal). Of the 5 patients (22.7%) who underwent urodynamic study, all had hyperactive detrussor and symptoms of urinary leakage could be controlled with anticholinergic medications. Clean intermittent catheterisation (CIC) was the commonest mode of bladder emptying as performed in 16 patients. One patient had suprabubic catheterisation and 5 patients voided by Valsalva manoeuvre. CIC were performed by the care giver of the patients.

Table 1: *Showing Demographic and Neurological Profile of Patients (n=22)*

Profile	Value
Age (mean, years) ± SD	40.41±15.18
Sex (male:female)	21:1
Surgery before admission	6(27.3%)
Time since injury (days, median)	34.5 (range 9-132)
Complete: incomplete	12:10
Tetraplegic: paraplegic	16:6
Motor score (mean) ± SD	45±24.97
Sensory score (mean) ± SD	157.50±69.53
FIM at admission (mean) ± SD	71.50±23.40
FIM at discharge (mean) ± SD	82.35±20.72

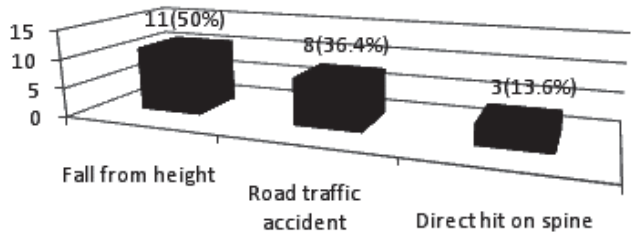

Fig 1- *Bar Chart Showing Causes of Traumatic SCI*

Discussion:

SCI has a great social and economic burden on society. Often the spinal injured subject is in the prime of life and the family is called upon to bear the expenses of hospitalisation and ongoing care. Hence, it is very important that we understand the characteristics of the disability to ascertain modifiable factors. The trends seen in this study are discussed below.

As evident from the results of this study, the largest number of patients was in the age range of 20–39 years constituting 10 patients (45.5%) closely followed by

Table 2: *Showing Motor and Sensory Levels*

Motor level	No. of cases (%)	Sensory level	No. of cases (%)
Cervical	**16 (72.7)**	Upper thoracic up to T6	**7 (31.8)**
Upper thoracic up to T6	**2 (9.1)**	Lower thoracic (T7-T12)	**4 (18.2)**
Lower thoracic (T7-T12)	**3 (13.6)**	Lumbar	**1 (4.5)**
Lumbar	**1 (4.5)**	Sacral Sparing	**10 (45.5)**

Table 3: *Showing Complications Observed at Admission*

Complications	No. of cases (%)
Spasticity	**16 (72.7%)**
UTI	**14 (63.6%)**
Pressure ulcer	**14 (63.6%)**
Orthostatic hypotension	**8 (36.4%)**
Shoulder pain	**1 (4.5%)**
Bladder stone	**1 (4.5%)**
Fistula (penoscrotal)	**1 (4.5%)**

that of 40–59 years accounting for 8 patients (36.4%). This is comparable to the age range observed in the study carried out by Chacko *et al*[1] wherein most patients were in the third or fourth decades of life based on a rural south Indian hospital based study. Agarwal *et al*[2] , in a hospital based study about demographic profile of SCI patients, also observed the age incidence of SCI in accordance. They also observed patients in 20-39 years age group comprising 41.5% while those in age group of 40-59 years constituting 35.3%.

Aetiology of SCI appears to have a major effect on the level and extent of neurological deficit as well as recovery[3]. Literature from developed countries reported that RTA accounts for the maximum number of spinal injuries[4] . Nogueira *at al*[5] in their study found wound by fire weapon as the commonest cause of traumatic SCI, accounting for 44.7% of the cases and RTA accounting 23.4% while fall representing only 17% of SCI patients.

There has been different observations regarding the commonest cause of SCI in developed /western countries as compared to our country. In the present study, fall from a height was observed to be the leading cause of injury accounting for 50% of the cases with RTA coming as next commonest cause accounting 36.4% of cases. Agarwal *et al*[2] also observed fall from height as the commonest cause of injury (58.9%) with RTA making the second commonest cause of injury (21.3%). However, Chhabra and Arora[6] observed RTA as the commonest cause of injury (45%) while fall from height accounted for 39.3%. This might be because of the population of SCI patients were mainly from in and around Delhi metropolitan city. And fall from height accounting for the second commonest cause of injury might be possible as patients from neighbouring states (of the country) might have been attending for better SCI management.

Previous studies have reported that the percentage of men affected was found to be more than that of women[3,7]. Comparable results were found in the present study.

Six patients (27.3%) in the present study underwent some form of surgery viz decompression ± internal fixation before admission in our ward. Reasons for undergoing surgery cited are ignorance, magnitude of the trauma including psychological as well as neurological impact following SCI, etc. Agarwal *et al*[2] also observed 25.6% of the patients underwent surgical intervention. However, the total number of patients in their series included non-traumatic SCI accounting for 12.6% and thus the % of patients of traumatic SCI who underwent surgical intervention should still be lower as compared to present study.

Kucukdeveci *et al*[8] in their study found that majority of the patients were paraplegics(72%). Agarwal *et al*[2] also reported higher incidence of paraplegics (63.8%). Chhabra and Arora[6] also reported 66.6% paraplegics among the SCI patients.This is quite contrast to the findings of our study where 16 cases (72.73%) were tetraplegics. The high incidence of tetraplegic may need attention of healthcare policy planners for more challenging rehabilitation strategy while framing rehabilitation intervention policy for SCI pateints in the region.

Agarwal *et al*[2] reported ASIA impairment scale (AIS) grade A, 33.9%; grade B, 18.6%; grade C, 17.8%; grade D, 30.5% amongst their SCI patients. Vijayakumar and Singh[9] reported 30% cases of AIS grade A, 25% cases of grade B, 10% cases of grade C and 35% cases of grade D. This is a bit in contrast to the findings of our study where there were (AIS) grade A, 54.55%; grade B, 0%; grade C, 18.18%; grade D, 27.27% cases. It

might be explained by the fact that the studies of Agarwal *et al*[2] and Vijayakumar and Singh[9] included patients of non traumatic SCI in 13% and 40% patients respectively. However, Chhabra and Arora[6] also observed 71% AIS grade A in their study. The high incidence of AIS grade A as observed by present study and Chhabra and Arora[6] might probably be indicative of need of acute trauma care services including transport from site of accident, for better neurological outcome of SCI patients in the country.

Vijaykumar and Singh[9] found 50% cases of traumatic SCI as modified dependence, 45% as no helper and 5% cases complete dependence as per FIM sub-grouping of the patients. However, the series, reported, consisted 60% traumatic SCI only out of the total 20 patients of SCI. In our study, the mean FIM score at time of admission was 71.50±23.40.

Kucukdeveci *et al*[8] in their study reported the most common complications observed during the rehabilitation period as urinary tract infections (73%), pressure ulcers (32%), orthostatic hypotension (17%), deep venous thrombosis (10%), and heterotopic ossification (7%). This is similar to our findings of UTI and pressure ulcer in 14 cases (63.64%). But spasticity was the most commonest complication observed in our study encountered in 16 cases (72.73%). And we also observed orthostatic hypotension in 8 (36.4%) of the patients. This may be attributable to high number of tetraplegic and high level paraplegic patients, both comprising 18 patients (81.8%) in the present study. Nogueira *et al*[5] also observed pressure sore complication in 42.5% patients. Vijaykumar and Singh[9] reported the commonest complication as neurogenic bladder which was seen in 70% of cases followed by spasticity in 60% of cases and pain in 45% of cases.

The limitations of the study are : small sample size, retrospective study and short duration of study.

Conclusion:

Majority of the traumatic SCI patients comprised ASIA grade A and tetraplegics were commoner than paraplegics. The commonest cause of injury observed was fall from height follwed by those due to road traffic accident. The commonest complications encountered in these pateints were spasticity, UTI and pressure ulcers.

Based on the above observations, a longitudinal future study with larger number of patients with traumatic spinal cord injury must be undertaken to investigate the long-term effects of the injury on functional status, community integration and quality of life measures. Other parameters, such as carers' stress and their quality of life might also be included.

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