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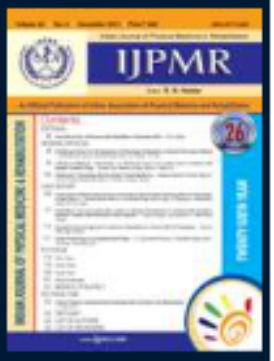

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Case Report

Brainstem Haemorrhage due to Autonomic Dysreflexia in a Person with C6 Tetraplegia

Swapna Patil[1], Raji Thomas[2]

Abstract

Autonomic dysreflexia is an important clinical complication occurring in patients with high levels of spinal cord injury. If untreated, the acute rise in blood pressure can cause end organ damage, including intracerebral haemorrhage. Though unusual, it can be fatal with large haemorrhages causing brain herniation syndromes. Here we report the case of a patient with C6 complete tetraplegia patient who developed brainstem haemorrhage during an episode of autonomic dysreflexia. The pathophysiology and treatment methods of this condition are discussed, highlighting the importance of preventive measures to avoid the same.

Key words: **Autonomic dysreflexia, spinal cord injury, cerebral haemorrhage, hypertensive encephalopathy.**

Introduction:

Autonomic dysreflexia is an acute medical emergency occurring in patients with spinal cord injury. Patients with tetraplegia and high paraplegia (at or above T6 level) are at risk of developing this complication[1]. In these patients, there is disconnection between the spinal sympathetic centres and the supraspinal sympathetic centres leading to loss of organised/ controlled sympathetic outflow below the level of the spinal lesion[2]. It is often triggered by non-specific stimuli below the level of lesion, bladder and bowel distension being the most common causes[1,2]. Clinical features frequently include sudden rise in blood pressure, headache, sweating, and flushing[2]. Early recognition and removal of the triggering factors leads to spontaneous resolution of the crisis. In case of inappropriate or delayed treatment, the sudden rise in blood pressure can cause end organ damage, including damage to the central nervous system leading to hypertensive encephalopathy or rarely intracerebral haemorrhage[1, 2].

Literature review shows few case reports of intracranial haemorrhage associated with autonomic dysreflexia. There are no previous reports of brainstem haemorrhage associated with this condition. Here we report the case of a patient with C6 tetraplegia who developed brainstem haemorrhage secondary to autonomic dysreflexia.

Case Report:

A 30-year-old male presentedwith history of weakness and loss of sensation in both lower limbs and upper limbs following fall from height in August 2010. He was diagnosed to have fracture of the C6 vertebral body with C6-C7 dislocation (Fig 1) and was managed with cervical traction in the Spinal Disorders Unit. Thereafter he was bedridden, developed pressure sores and was on indwelling urethral catheter.

Eight months after the event, he was referred to us for rehabilitation. On examination he was found to have C6 complete tetraplegia with pressure sores and chronic collapse of the left lung secondary to a mucus plug in the bronchial tree.The pressure sores were managed conservatively. Left lung collapse was managed with chest physiotherapy. With 6 weeks of rehabilitation,he was discharged to be partially independent from a wheelchair and was on an indwelling catheter (clean

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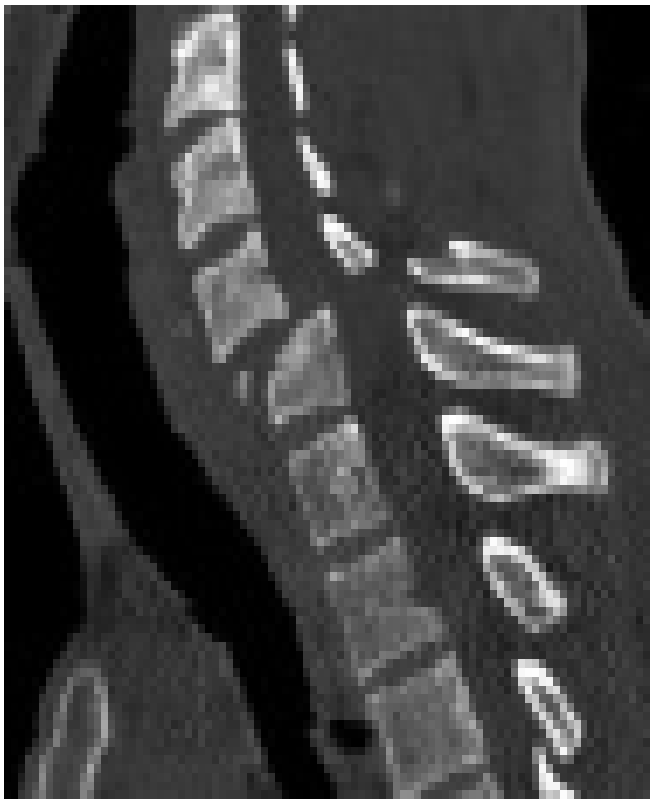

Fig 1: CT Cervical Spine Sagittal Image Showing Fracture of the Spinous Process of C6 Vertebra, Anterolisthesis of C6 over C7 and Compression Fracture of Anterosuperior Border of C7

intermittent catherisation was not considered in view of poor hand functions), engaging in a meaningful vocation. Two months following discharge, he again presented withpounding headache,right facial paralysis and double vision of three days duration. He gave history of an episode of bladder distension with an increase in blood pressure to 180/110 mm Hg associated with the onset of these symptoms. The blood pressure hadcome down over few hours after bladder evacuation. On examination,he had right sided partial ptosis, right lateral rectus palsy, right sided gaze evoked nystagmus, right sided LMN facial palsy and impaired gag reflex. MRI brain showed a 10x12x15 mm lesion situated inferomedial to the right middle cerebellar peduncle and posterolateral to the medulla. This was isointense to the gray matter and non-enhancing. Marked blooming was seen on SWI images (Fig 2). Features were suggestive of subacute haematoma.

During the hospital stay, he continued to have episodic headaches which were associated with sudden rise in blood pressure. He was started on amlodipine 2.5 mg once daily with which blood pressure normalised. After the acute phase settled he was continued on the

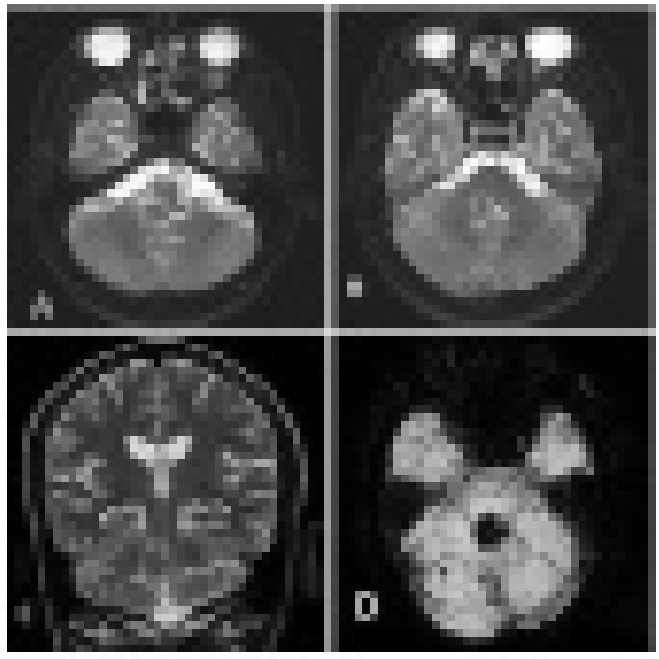

Fig 2 : MRI Brain T2 Axial (A and B) and Coronal (C) Images Showing Isointense Lesion with Surrounding Area of Hyperintensity Inferomedial to the Right Middle Cerebellar Peduncleand Posterolateral to the Medulla. SWI (D) Image Shows Marked Blooming of the Lesion

rehabilitation programme.Later amlodipine was stopped and suprapubic catheterisation was done. There were no further episodes of autonomic dysreflexia.

Discussion:

Autonomic dysreflexia (AD) was first observed by Anthony Bowlby in 1890, and described by Guttmann and Whitteridge in 1947. It is seen in patients with cervical or high thoracic cord injury (at or above T6 level below which the main sympathetic outflow exits from the spinal cord), occurring in up to 50 - 90% of these patients. AD increases with ascending level and completeness of SCI/ injury severity. It is 3 times commoner after complete injuries than after incomplete injuries[1-4].

AD after spinal cord injury is due to changes in the spinal and peripheral autonomic circuits, occurring during the acute and chronic stages of SCI. The mechanisms involved include loss of supraspinal inhibitory control due to destruction of the descending vasomotor pathways, plasticity occurring within the spinal cord mainly at the level of the spinal sympathetic neurons and the primary afferents and changes in the sensitivity of peripheral adrenergic receptors[2-4]. It usually develops weeks to months after the SCI, but can occur as early as 4 days after severe cervical cord injuries[2, 4, 5].

Noxious and non-noxious stimuli can trigger this condition, with common stimuli being bowel and bladder distension, spasms, and pressure sores. Other triggers include urethral catheterisation, bladder percussion, urinary tract infections, cystoscopy, cystometry, and electrical stimulation of muscles[2-5].

AD is characterised by episodes of extreme hypertension (systolic blood pressure up to 300 mmHg). As spinal cord injury patients usually have low systolic BP (90 to 110 mmHg range or 15 to 20 mmHg lower than normal people), an increase in blood pressure by 20 to 40 mmHg above the baseline can also be considered as a feature of AD, if associated with other features. Intensity of these episodes varies from being asymptomatic, or associated with mild discomfort and headache to a life threatening hypertensive emergency/ encephalopathy[1-6].

Clinical features include hypertension, bradycardia, pounding headache, increased spasticity, sweating, blurred vision, nasal congestion, cutis anserine, piloerection, upper body flushing, and apprehension. If untreated, these episodes can cause intracranial haemorrhage with risk of brain herniation, cardiac complications, retinal detachment, seizures and death[1-6].

Previous reported cases of intracerebral haemorrhage following autonomic dysreflexia were in the left putamen[7], left basal ganglia and thalamus leading to death[5], left occipital region[8], right thalamus[9], cerebellum[10] and right putamen with extrinsic compression of the lateral ventricle[1]. Massive right cerebral haemorrhage with rupture into the lateral, third and fourth ventricles with bilateral uncal, trans-tentorial and cerebellar tonsillar herniation has also been reported[6].

Our patient had C6 complete tetraplegia and he developed features of AD related to bladder distension 10 months after the injury. During this episode he had sudden increase in blood pressure and subsequently brainstem haematoma. A literature search could not find any previous reports of brainstem haemorrhage following autonomic dysreflexia. The LMN facial palsy could have resulted from involvement of the facial nucleus in the pons;due to extension from the dorsolateral medullary lesion. This episode subsided with bladder evacuation, but produced new neurological deficits in addition to the old deficits. On follow-up there was improvement in the diplopia and ptosis but facial palsy persisted.

Conclusions:

Brainstem haemorrhage, though a rare complication of autonomic dysreflexia, due to its potential morbidity and mortality becomes significant in patients with spinal cord injury. Preventive measures, early diagnosis and adequate treatment would avoid life threatening complications related to AD.

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Original Article

A Study of Effects of Intervention of Botulinum Toxin- A on Lower Limb in Children with Spastic Cerebral Palsy

Kumar Raj[1], Wadhwa Sanjay[2], Singh U[3], Yadav SL[4]

Abstract

Objective: Analysis of clinical gait pattern, change in spasticity and range of motion (ROM) in cerebral palsy patient (CP) with spastic lower limb muscle after injecting botulinum toxin- A.

Study Design: Prospective study

Methods: 28 children (18 male and 10 female) with spastic CP had problems in normal walking, aged 2–9 years (mean age 4.65 years), consecutively treated in the PMR department over a 2-year period, were prospectively followed-up and clinically assessed pre- and post-treatment (at 2 weeks and 2 months) both objectively and subjectively. Objective assessment included gait parameters -- stride length, cadence, velocity, step length, base of support; active and passive range of motion (ROM), (measured by goniometry) and spasticity on modified Ashworth scale. Subjective assessment was done by asking questionnaire in terms of comfort, ease of care, perineal hygiene, walking. Injections were given using clinical palpatory method on OPD basis. All patients received botulinum toxin-A injections, followed with exercises and activities and orthosis as needed.

Results: Significant improvement was achieved for spasticity reduction in gastrocnemius ($p< 0.001$), hamstring and adductor ($p=0.050$), ankle AROM & PROM ($p< 0.001$), active knee extension ($p=0.009$), popliteal angle ($p=0.015$) and percentage left and right foot contact ($p< 0.001$), whereas non-significant change was observed in step length, cadence, velocity, stride length, and base of support. Parents felt subjective improvement in most of the cases (>90%).

Conclusions: Botulinum toxin- A injection is effective in the treatment of spastic lower limb muscles for equinus/ crouching/scissoring gait in cerebral palsy children. The treatment was feasible and easily implemented. Botulinum toxin- A injections were well tolerated, yielded no serious treatment-related adverse events.

Key words: **Botulinum toxin-A, spastic cerebral palsy.**

Introduction:

Cerebral palsy (CP), a group of permanent, non-progressive congenital neurological disorders, is characterised by the resulting effects on movement and posture. Graham argued that the basic definition of CP should be extended to include the progressive nature of the musculoskeletal pathology. Motor impairments that result from the many neurological deficits in CP include neuromuscular and musculoskeletal problems of spasticity, dystonia, muscle contractures, bony deformities, incoordination, loss of selective motor control, and weakness. The stiffness of the lower limbs often deteriorates during development results in defective walking patterns like scissoring gait due to hip adductor tightness, crouched gait due to hamstring tightness and toe walking due to calf muscle tightness. Therefore, antispasticity treatment , plays an important role in treating the child with CP. There are numerous treatment options available for hypertonicity management which is characteristic feature of CP, including physical and occupational therapy , orthosis , oral medications like baclofen, tizanidine, dantrolene etc, chemodenervation tendon lengthening, and dorsal rhizotomy. Thus a stepped up management protocol is adopted, beginning with the more conservating options

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and reserving surgical option for older children and those cases where the stiffness and progressive deformities continues to hamper rehabilitation. Chemodenervation is commonly done by botulinum toxin-A (BTX-A), phenol and alcohol. BTX-A is potent neurotoxin produced by anaerobic bacterum *Clostridium botulinum* with serotype A. It causes chemodenervation and muscle relaxation by inhibiting acetylcholine release into the synaptic cleft thus blocking the neuromuscular junction. The denervation temporarily reduces muscle tone and provides an opportunity to effect changes in motor learning and cortical motor organisation. BTX-A, when compared with phenol for adductor spasticity, improves cadence, step length, and velocity during gait . Anatomic localisation of target muscles using palpation was the most common technique and injection can be done quickly; and pain was not commonly reported as an important factor after injection. To date, most randomised controlled trials on the effects of BTX-A injections have focused on treatment of the lower extremities, including the ankle flexors, knee flexors, and hip adductors in children with spastic diplegia and hemiplegia. The two most common impairments studied are spasticity and limitations in range of motion (ROM). The level of function has been assessed by observing gait or reviewing the results of motor and functional outcome scales. The most commonly used tool to assess spasticity is the modified Ashworth scale (MAS). Multiple randomised controlled trials have demonstrated reduced MAS scores after injection of BTX-A in ankle flexors, hamstrings, and hip adductors. Outcome measures were not similar, and different in different studies. This study was conceptualised to evaluate the effects of BTX-A injection on lower limb muscles especially in terms of clinical gait pattern, spasticity and ROM.

Materials and Methods

Type of study – Prospective study.

Study population - 34 spastic CP patients, 2 to 9 years aged of either sex, who attended the outpatient department (OPD) of Physical Medicine & Rehabilitation (PMR), at the All India Institute of Medical Sciences (AIIMS), New Delhi between 2009 and 2011. Patient selection was done with the following criteria:

Inclusion criteria:

1. Spastic cerebral palsy patients with involvement of lower extremity muscles e.g. adductor alone, calf alone, or adductor with medial hamstring or adductor with calf muscle (gastrocnemius) or all three muscle group, having spasticity interfering in gait pattern e.g. scissoring gait due to adductors, crouched gait due to hamstring , toe walking due to calf muscles.
2. Age 2 to 9 years,
3. Willingness of the parents/ guardians to participate.

Exclusion criteria:

1. Fixed joint contractures or deformities.
2. Bleeding disorders.
3. Previous treatment with BTX-A within 6 months.
4. Concomitant treatment with phenol, alcohol or any neurolytic procedures.
5. Known allergy to BTX-A.
6. Spasticity not interfering with activity of daily living or walking.
7. Cognitive dysfunction to such an extent that the patient will not be able to cooperate or follow instructions.
8. Patient on aminoglycosides.

BTX-A, is approved by US FDA, for the treatment of spasticity in cerebral palsy patient. The ethical clearance was obtained from the institutional ethical committee prior to the commencement of this study. The patients were explained about the study and their written consent was taken. Then their clinical assessment (history and physical examination) was being done. There was at least one week pre study observation period (for conservative management). Before injecting BTX-A, assessment was done in following way:

Assessment of gait parameters:

a) Stride length
b) Cadence
c) Velocity
d) Step length
e) Base of support
f) Number of falls if any

Active ROM, passive ROM (measured by goniometry) and grading of muscle spasticity on MAS and adductor tone rating were noted.

Subjective assessment: Questionnaire in terms of comfort (feeling better than earlier), ease of care, perineal hygiene, walking. Answer was taken as yes/ no.

After that two post injection follow-up at 2 weeks and 2 months were done. At each follow-up outcome measures of both objective and subjective were noted.

No change in antispastic medication was done during the course of the study.

Modified Ashworth scale 15 (for spasticity grading of hamstring and gastrocnemius):

0: no increase in muscle tone

1: slight increase in muscle tone, manifested by a catch and release or by minimal resistance at the end of the ROM when the affected part(s) is moved in flexion or extension

1+: slight increase in muscle tone, manifested by a catch, followed by minimal resistance throughout the remainder (less than half) of the ROM

2: more marked increase in muscle tone through most of the ROM, but affected part(s) easily moved

3: considerable increase in muscle tone, passive movement difficult

4: affected part(s) rigid in flexion or extension

Adductor tone rating (for adductor spasticity assessment):

0: No increase in tone.

1: Increased tone, hips easily abducted to 45º by one person.

2: Hips abducted to 45º by one person with mild effort.

3: Hips abducted to 45º by one person with moderate effort.

4: Two people required to abduct the hips to 45º.

Intervention:

BTX-A is marketed as Botox® (Allergan, Inc.), Dysport® (Ipsen Limited), a Chinese formulation, Hengli (Lanzhou Institute of Biologic Products), and Xeomin ® (Merz Pharmaceuticals), Botox® was used (Allergan, Inc.) in this study in all patients because it was easily and widely available.

Botox® injection were administered under complete aseptic precaution. Injection site of muscle was chosen as per anatomical landmark near the site of motor innervations by clinical palpatory method. Following injections, the muscles was massaged and subjected to ROM exercise a few times. Patients were observed for 1 hour, post injection to evaluate possible adverse events (pain, bleeding, rash, allergic reaction and other concerns) if any. Patients were instructed to contact us directly over mobile phone with concerns regarding possible side-effects if any. Patients and their caregivers were reviewed 2 weeks and 2 months following each injection protocol for assessment.

Dosage: 4 U/kg per muscle with maximum of 50 U per injection site. Dilution was done by dissolving 100 U in 2 ml (50 U/ml) 0.9% normal saline. Maximum dose given was 12U/ kg total body weight for different muscles injected in a sitting.

Injection procedure: It was done on OPD basis. Attention diversion (like songs on mobile, engaging the child in different talks e.g. stories, etc) of the child was done while giving injections to alleviate fear of injection and then injection was given under complete aseptic and sterile condition. Injections were given by 1ml tuberculin syringe (26 gauze needle). Localisation technique was solely on anatomical basis where muscle belly is most prominent i.e. at the site of motor point and localisation of the injecting needle through fascia of the target muscle. In some very anxious children, sedation with syrup promethazine hydrochloride or triclofos was given.

After BTX-A treatment, the children continued participating in routine physical therapy throughout the study period as doing earlier. Children were provided knee ankle foot orthosis (KAFO) with or without abductor bar, ankle foot orthosis (AFO).

PROM at ankle with the knee in maximum extension was evaluated, and the foot was held in supinated position to diminish subtalar motion and mid foot dorsiflexion. The heel was positioned in the palm with the forefoot on my forearm, ensuring that the calcaneus was in neutral and the foot was aligned with the tibia. Maximum passive dorsiflexion was measured by using manual goniometer.

AROM at ankle was obtained from the flexor withdrawal reflex to determine any change in ROM during treatment. It was ensured that foot did not deviate into inversion or eversion. Maximum active dorsiflexion was measured with manual goniometer.

Popliteal angle was measured between the femur and the tibia as measured after flexing the hip to 90 degrees.

Active knee extension was measured as angle between longitudinal axis of the femur and tibia .

Assessment of gait parameters:

Stride length, base of support and percentage length of foot contact were measured by direct floor technique, [For simplification and easy quantification of foot contact part of physician rating scale (PRS) in busy OPD setting, measurement of the percentage of foot contact length antero-posteriorly from heel to toe was done], by applying chalk powder on the feet of child. Measurement of base of support was obtained by calculation of perpendicular distance between the most medial point of the inner border of each foot. Stride length was measured as distance between 2 successive placements of the same foot.

For cadence, velocity, step length child was asked to walk over a measured 10 metre distance. Out of 3 best measurements median value was taken.

Number of steps and time taken to cover the 10 metre distance is noted and then cadence, velocity and step length were calculated arithmetically.

Statistical analysis:

This was a prospective study with two follow-up at 2-week and 2-month after ascertaining baseline values. Total sample size was twenty-eight (28). For descriptive studies, various statistical parameters like the mean, median, standard deviation and range were used for determining continuous variables and frequency, percentage and range were used for determining the categorical variables. Besides descriptive statistics, the comparison over period of time was done by applying repeated measures analysis followed by post hoc comparison by Bonferroni method. Besides this where data were not distributed normally, Friedman test was applied. The comparison between pre and post injection follow-up was done by applying Mc Nemar test for the follow-up at 2-week and 2-month.

Log transformation was also applied where the data was not distributed normally and sample size was reasonable. P - value <0.05 was considered as significant.

Results

A total of 28 patients (18 diplegic, 10 hemiplegic), 18 male, 10 female, 2-9 years old (mean age 4.66 yrs) completed the study. Out of 18 diplegic, 8 patients were given injection in both gastrocnemius, 4 patients in bilateral (b/l) adductor and gastrocnemius, 3 in b/l adductor and hamstring, 2 patients in b/l hamstring and gastrocnemius, 1 patients in b/l hamstring and left gastrocnemius. All hemiplegics (7 left and 3 right hemiplegics) were given BTX-A injection in respective gastrocnemius only. Total 69 muscles (43, gastrocnemius, 14 adductors and 12 hamstrings) were injected(Fig 1 & Tables 1-4).

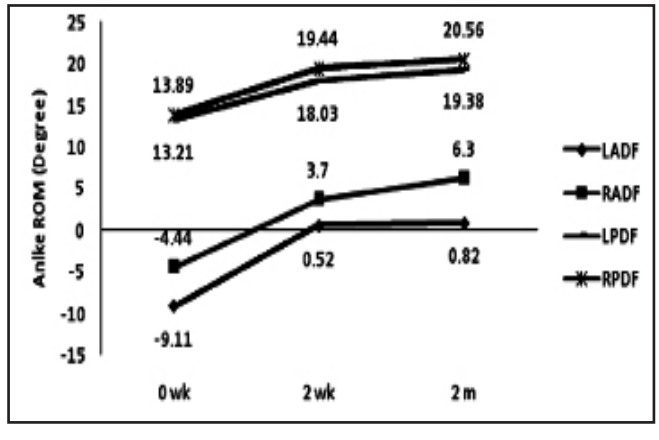

Fig1: ROM at Ankle (n= 28)

(LADF&RADF - left & right ankle active dorsiflexion; LPDF & RPDF- left & right ankle passive dorsiflexion dorsiflexion) p<.001

Table 1: ROM at Knee

AROM	ROM 0 Week Median (minimum, maximum)	ROM 2Weeks Median (minimum, maximum)	ROM 2 month Median (minimum, maximum)	P
LAKE (n=6)	-17.5(-30,-5)	-7.5 (-20,0)	-5(-20,0)	.009
RAKE (n=6)	-17.5(-30,-5)	-7.5(-20,0)	-5(-20,0)	.009
LPA (n=6)	70(55,70)	60(55,60)	55(50,60)	0.015
RPA (n=6)	70(55,70)	60(55-60)	55(50,60)	0.015

(LAKE -Left active knee extension); (RAKE - Right active knee extension) (LPA-Left popliteal angle); (RPA- Right popliteal angle).

Table 2: Gastrocnemius Spasticity

Muscle	Mean ± SD/0 week	Mean ± SD/ 2 week	Mean ± SD / 2months	P
LG (n=24)	2.92 ± O.282	2.25 ± 0.442	2.08 ± 0.408	<.001
RG (n=19)	2.95 ± 0.229	2.26 ± 0.452	2 ± 0.471	<.001

(LF- left gastrocnemius, RG- left gastrocnemius)

Table 3: Hamstring and Adductor Spasticity

MUSCLE	Median (minimum, maximum)/0 week	Median (minimum, maximum)/2weeks	Median (minimum, maximum)/2months	P
LH (n=6)	2(1-3)	1(1-2)	1(1-2)	0.050
RH (n=6)	2(1-3)	1(1-2)	1(1-2)	0.050
LA (n=7)	2(1-3)	1(1-2)	1(1-2)	0.050
RA (n=7)	2(1-3)	1(1-2)	1(1-2)	0.050

(LH-left hamstring, RH- right hamstring, LA-left adductor, RA-right adductor)

Table 4: Gait Parameters

GAIT P	Mean±SD/0week	Mean±SD/ 2weeks	Mean±SD /2months	P
STR L (n=28)	48.16±12.67	48.45±11.33	50.64±10.63	.075
CAD (n=28)	95.16±43.88	94.47±41.46	101.22±36.50	0.210
VEL (n=28)	26.54±16.45	26.76±14.19	28.43±12.75	0.242
STEPL(n=28)	25.75±7.09	26.29±6.41	27.77±5.89	0.033
BS (n=28)	9.43±9.63	10.27±8.92	10.41±7.93	0.134
% LFC(n=28)	56.82±21.13	78.05±19.01	81.85±16.03	<.001
%RFC(n=28)	61.61±24.16	75.75±8.79	84.54±4.21	<.001

(STR L: Stride length, CAD: Cadence, VEL: Velocity, STEP L: step length, BS: Base of support LFC and RFC: left & right foot contact)

Subjective improvement: 27 out of 28 (96.4%) parents of patients felt comfort; 28 out of 28 (100%) parents felt improvent in ease of care ; 26 out of 28 (92.9%) parents of patients felt there is improvement in walking ; 7 out of 28 (25%) parents of patients were asked about improvent in pereneal hygeine. All 7(100%) parents of patients reported subjective improvement in perineal hygiene at 2 week which remained till 2 months.

Side-effects: (1) Transient localised weakness-in one patient (BTX-A was injected in bilateral hamstring) (2) Flu like symptoms (fever) were noticed in two patients one day after injection which responded with in 2 days after taking syrup paracetamol.

Discussion:

Treatment with Botox® was effective in reduction of spasticity in the gastrocnemius, adductor and hamstring, increased active and passive ROM of the ankle joint, active knee extension confirming the findings of pre¬vious studies as described and discussed below.

Koman *et al*[1] showed that BTX-A treatment of the medial and lateral gastrocnemius muscles can reduce spasticity and improve gait and positioning in children with progressive dynamic equinovarus or equinovalgus foot/ankle deformities that were unresponsive to bracing. We also tried to correct these deformities with

bracing, physical and occupational therapy but got better result with intervention of BTX-A injection.

In a prospective, 3-month, randomised, double-blind clinical study by Koman *et al* [2] involving 114 children with CP and dynamic equinus foot deformity, patients in the Botox® group demonstrated improved ankle ROM, gait function and a 20% reduction in motor evoked potential (MEP) of the gastrocnemius muscle. The authors rated the predictability of improved gait function as low which is similar to our study.

Wissel *et al*[3] found a significant improvement in Ashworth score ($p<0.001$), knee ROM ($p<0.01$) (passive 15°, active 20°) after BTX-A injections in multiple muscle groups, including the hamstrings, hip adductors, and ankle flexors. While gait analysis revealed significant increase in gait velocity ($p<0.01$) and stride-length ($p<0.001$) over baseline. In our study we did not get such improvement in velocity or stride. The reason could be less number of patients had injection in adductor and hamstring muscles

Fazzi *et al* [4] observed a reduction in spasticity and an increase in passive joint mobility and scores on scales and tests intended to assess function. These improvements are reflected in the gait pattern (PRS), selective motor control in foot dorsiflexion, and the acquisition of new motor abilities.

Furthermore, the muscular relaxation achieved following application of BTX-A allows muscle-stretching exercises to be performed more easily and the child to learn to use and strengthen opposing muscles. For most children BTX-A helps delay shortening of the muscles. Specific therapeutic exercise treatment and orthosis can help the small percentage of children in whom there is evidence of prolonged muscle stretching. These are clear arguments for intensifying exercise therapy after the application of BTX-A to achieve better results in the long term as our patients did and could be able to tolerate easily.

Although the effects of BTX-A are seen primarily in a reduction of hypertonia, the changes in tone can improve the child's balance, strength, motor control and fixed contractures . As the child develops, the spastic muscles fail to grow as quickly as the neighbouring structures, and dynamic structures are transformed into fixed contractures. Relaxation of spastic muscles allows these to stretch, encourages their growth and prevents contractures. These improvements lead to functional gain.

It has been stressed that assessment of functional improvement is more important than assessment of the reduction in muscle tone or the increase in the ROM of the joint . The physiological and mechanical effects of BTX-A treatment are genuine and measurable in patients with spastic diplegia. However, these effects may not trigger a major change in function or change in the patient's or his/her parents' perception of function and thus not be recorded as a significant improvement in the patient's family and social life . But in our study there is great improvement in parents' perception of function; it may be due to early injection BTX-A (mean age 4.65years) as well as less strictness of these improvement criteria.

We also observed an improvement on the foot contact component related to PRS scale after treatment; the beneficial effect was sustained until the end of the study. The scale sub items gait pattern and especially percentage of left and right foot contact, which are the two elements that are most closely related to spasticity of the gastrocnemius, improved significantly with BTX-A. Gait speed, the item with the greatest functional value on the scale, also improved but it was not significant statistically.

It was noticed that the non-significant improvement in gait speed, step as well as stride length is corroborating the findings in most of the studies done in CP patient. It may be due to more number of patients affected with gastrocnemius spasticity and less of adductor and hamstrings. Improvements in gait speed, step as well as stride length may also be contributed due to increased growth of the child especially in multiple and prolonged follow-up. On the contrary these improvements were noted significant in most of the studies done in post stroke spasticity as they tried better use to motor relearning because they knew their normal premorbid status.

Adductor injection may reduce scissoring, and knee flexor injection may aid knee straightening and improve standing ability. One class I study using BTX-A injection ($n=61$) into the adductors and medial hamstrings of CP patient showed an average improvement in knee-to-knee distance(fast catch) of about 9 cm ($p < 0.002$) and decrease in adductor spasticity on modified Ashworth scale of 2 ($p < 0.001$) . Significant improvement noted both at 4 and 12 weeks. Two small open-label studies (class IV) found modest improvement in either gait kinematics or hamstring length with BTX-A injection into the hamstrings.

Corry *et al* [5] injected BTX-A into 17 hamstrings in CP patients (mean age 7.2; Range 4 -11yr). In this study by Corry *et al* [5] showed the median MAS score for hamstring were equally decreased. Spasticity at 0 week was 1(1-2) which decreased 1(0-2) at 2 weeks and 1(0-1) at 3 months. This change was not significant. In our study spasticity decreased by grade 1 in almost all patients but this was not significant statistically as only 6 children had hamstring spasticity.

Mean popliteal angle decreased by 16° at 2 weeks from mean±S.D of 64°(12.8) to 48°(11.3); and 17° at 3 months mean±S.D of 64°(12.8) to 47°(7.3); (n=10). It correlates with our finding as median decrease was 10˚ at 2 weeks and 15˚at 2 months from 70˚ at baseline which is significant. The additional 5˚decrease in mean popliteal angle can be attributed to muscular relaxation achieved following application of BTX-A allows muscle-stretching exercises more actively and easily. Our study noted improvement in the active knee extension of 10˚ at 2 weeks and 12.5˚ at 2 months from median baseline of -17.5˚, almost similar to Corry'*et al* [5] study; this improvement was also noted qualitatively in stance phase. This means static improvement in active knee extension might be reflected dynamically in stance phase of the gait cycle.

So it is clear from various studies that BTX-A has a major effect on the dynamic spastic component but only limited effect on passive ROM, since the latter depends of resistance produced by muscle or joint connective tissue . In our study also more change is noted in AROM compared PROM, because some of our patients already had dorsiflexion passive ROM at ankle was ≥ 20˚. We also believe that the increased effect of BTX-A at 2 months may also be related to the results of more aggressive therapeutic exercise .

In a recent study Carlos *et al*[6] evaluated 20 children with spastic diplegic CP children. All the patients received injections in the gastrocnemius and soleus, and 15 received injections in the adductors. As like others they found decreased spasticity in gastrocnemius and adductors, decreased heel-ground distance , improved ambulation and equinus gait pattern, PRS ($p<0.001$). Gait speed also changed significantly as a result of the use of BTX-A ($p<0.05$).

Gait velocity in our study did not increase significantly, it may be explained by less number of adductor and hamstring injection in our study. The heel-ground distance is significantly decreased which may be correlated with increased percentage contact of foot to the floor of our study.

Degelaen *M et al* [7] recently suggested, botulinum toxin injection in lower limb spastic muscles leads to changes in motor planning, including thorough interference with trunk stability, but a combination of therapies (orthosis and physical therapy) is needed in order to learn new motor strategies.

We assessed children from 2 to 9 years of age (mean 4.65 years), and our initial results were significant. With younger children we might have been able to maintain the functional gains because the motor pattern of very young children provides greater scope for better development and recovery (a younger child has greater potential than an older child for increasing the plasticity of the central nervous system).

A study by Wissel J *et al* [3] revealed that of 33, 16 (48%) subjects reported improvement. Baker *et al*[8] study showed more than half of parents reported good or minimal improvement. In our study parents reported subjective improvement in more than 90% of all cases that might be explained by younger children (mean age 4.44 years) and less strict subjective criteria followed in this assessment.

Most of the studies did not report any serious adverse; the rare side-effects were mild and transient with CP . In our study only one patient had fever. This is revealing that if BTX-A is injected properly, adverse effects are very minimal.

Conclusions:

BTX-A injection of the lower limb muscles was effective in the treatment of spastic equinus/crouching/scissoring in patients with CP. There was significant reduction in spasticity, popliteal angle and increase in ROM at ankle, active extension of knee. Percentage length of foot contact is increased. But evidence for improved walking is not so convincing in terms of gait velocity, stride length and cadence. Study limitations were short follow up, lack of control group and not verified the extent of physical exercise and orthosis wearing time.

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Editorial

International Day of Persons with Disabilities, 3 December 2015

The International Day of Persons with Disabilities (IDPD) has been commemorated since 1992 to promote awareness and mobilise support for critical issues relating to the inclusion of persons with disabilities in society and development. The Day works to promote action to raise awareness about disability issues and draw attention to the benefits of an inclusive and accessible society for all.

Theme for IDPD (2015: Inclusion matters: access and empowerment for people of all abilities)

The estimated one billion people living with disabilities worldwide face many barriers to inclusion in many key aspects of society. As a result, people with disabilities do not enjoy access to society on an equal basis with others, which includes areas of transportation, employment, and education as well as social and political participation. The right to participate in public life is essential to create stable democracies, active citizenship and reduce inequalities in society.

Persons with disabilities must be able to fulfil their role in society and participate on an equal basis with others. It is important to focus on the ability and not on the disability of an individual. Often, the societal image of persons with disabilities is impacted by attitudes based on stigma and discrimination, as well as archaic ideas about disability and persons with disabilities that are often the greatest barrier to their full and equal participation in society and development on an equal basis with others. It is important to note that disability is part of the human condition, and that all of us either are or will become disabled to one degree or another during the course of our lives.

By promoting empowerment, real opportunities for people are created. This enhances their own capacities and supports them in setting their own priorities. Empowerment involves investing in people - in jobs, health, nutrition, education, and social protection. When people are empowered they are better prepared to take advantage of opportunities, they become agents of change and can more readily embrace their civic responsibilities.

Sub-themes for IDPD 2015:

- **Making cities inclusive and accessible for all**

It is estimated that by 2050, 66% of the world's population will be living in cities.Importantly, the New Urban Agenda must ensure that future cities, towns and basic urban infrastructures and services are more environmentally accessible, user-friendly and inclusive of all people's needs, including persons with disabilities. The International Day will be used to discuss and present some best practices of inclusive urbanisation.

- **Improving disability data and statistics**

The lack of data and information on disability and the situation of persons with disabilities at the national level contribute to the invisibility of persons with disabilities in official statistics. This presents a major obstacle to achieving development planning and implementation that is inclusive of persons with disabilities. In particular, to be internationally comparable, data should be collected in line with international standards. Data collected can be used the implementation and monitoring of internationally agreed development goals for persons with disabilities.

The International Day will be used to highlight measures to strengthen national capacities to improve and mainstream disability data collection, based on existing good practices. The Day will also be used to highlight challenges and map out strategies to involve persons with disabilities and their organisations in disability data and statistics collection and dissemination.

- **Including persons with invisible disabilities in society and development**

Persons with mental and psychosocial disabilities represent a significant proportion of the world's population. Millions of people worldwide have mental health conditions and an estimated one in four people globally will experience a mental health condition in their lifetime. Almost one million people die due to suicide every year, and it is the third leading cause of death among young people. Persons with mental and psychosocial disabilities often face stigma and discrimination, as well as experience high levels of physical and sexual abuse that occur in a range of settings, including prisons, hospitals and homes. Persons with other invisible disabilities, such as persons with hearing impairments, are also at risk of exclusion from mainstream activities, education or social activities.

The International Day can be used to draw attention on the situation of persons with invisible disabilities, such as mental health and psychosocial disabilities, intellectual disabilities, as well as hearing impairments. The Day can be used to identify good practices of integrative and inclusive education, to organise social activities and awareness raising initiatives, as well as highlight good practices and make recommendations.

R.N. Haldar

Case Report

Hitchhiker's Toe and Lumbar Disc Lesion: A Strange Coincidence in a Patient with Idiopathic Dystonia Treated with Botulinum Toxin Injection

Ayyoub Baqer[1], Jai Shanthini S[2], Wael Abdul Gawad [3]

Abstract

A 40 years old female with generalised dystonia and chronic low back pain was injected with botulinum toxin injection to extensor hallucis longus(EHL) for management of Hitchhiker's toe. The patient benefited functionally but later developed EHL and extensor digitorum (ED) weakness. Further workup revealed degenerative L4/5 disc lesion causing L5 root affection as the cause for the weakness. This coincidental occurrence of EHL and ED weakness due to L4/5 disc lesion in a patient with generalised dystonia , following botox injection to EHL for management of Hitch hiker's toe, has not been reported before.

Key words: **Dystonia, Hitch hiker's toe, botulinum toxin injection.**

Introduction:

Hitchhiker's toe has been defined as an apparent spontaneous extensor plantar response, without fanning of the toes. Other terminologies include striatal toe[1] or persistent extension of great toe or spontaneous Babinski sign or overactivity of the extensor hallucis longus (EHL). It occurs in patients suffering from sroke and dystonic syndromes. It causes painful spasms and gait disturbances including abnormal push-off and forward propulsion towards final stance[2]. Botulinum toxin injection is effective in the management of Hitchhiker's toe seen in patients with dystonic syndromes. Dystonia could also result in early spinal degeneration[3]. In this report we present a patient with idiopathic dystonia who was treated with botulinum toxin A injection to extensor hallucis longus muscle for the management of Hitchhiker's toe and was later found to have EHL and ED weakness due to degenerative lumbar disc lesion.

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Case Report:

The patient was a 40 years old Caucasian female who suffered from generalised dystonia. She was born by caesarian section and was one of twins. She had completed secondary school and was employed in a clerical job. Her main complaint was painful spasms involving the trunk and lower limbs resulting in recurrent ankle injuries since childhood for which she had consulted neurologist for the first time at the age of 35 years. CT brain done twice was normal. Work up for secondary dystonia including serum ceruloplasmin was negative. She improved symptomatically with trihexyphenydyl (Artane) 2 mg twice daily. Subsequently the dystonic movements had decreased.

However she continued to have difficulty in walking due to bilateral persistent extension of great toes and her shoe uppers were getting torn frequently. She also complained of chronic low back pain of 5 years duration and spasms in both hands while writing suggestive of writer's cramps.

On examination, patient was found to have mild dysarthria; muscle power was normal in both upper limbs and in bilateral hips, knees and ankles. Dystonic movements were observed in both feet in the form of persistent extension of bilateral great toes and mild

inversion of left foot which was exacerbated during walking. The great toe hyperextension was functionally disabling for the patient and hence it was decided to treat it with botulinum toxin injection. Injection botulinum toxin A was injected at therapeutic dose (Dysport 100 units) to bilateral extensor hallucis longus muscle after localising the muscle with ultrasonogram.

Patient was reviewed four weeks after the injection. We used the following scale for Shoe difficulties and pain : 0- absent 1- mild, 2- severe and 4 point scale for EHL overactivity as 0- no overactivity, 1- triggered by walking, 2- triggered by standing up, 3- triggered by any stimulation. The functional benefits after the injection in our patient were as follows:

On both sides, there was no more pain or damage to shoe upper. Pain and shoe difficulties scale was 2 preinjection and 0 post injection. On the right side , the EHL overactivity was still present but it was lesser - pre injection scale was 3 and post injection scale 1.

However on the left side, there was weakness of EHL as well as extensor digitorum (ED). She also complained of radiating pain to the left lower limb and exacerbation of low back pain since two weeks . There was tenderness in the lumbar spines L4 and L5 with painful restriction of lumbar flexion. On left side, there was weakness of extensors of all toes and diminished sensation in L5 dermatome. MRI confirmed L4/5 disc bulge with caudal migration and compression of bilateral L5 roots (Figs1&2). Nerve conduction study of lower limbs was normal and patient did not agree for needle EMG. Neurosurgeon advised surgery for the L4/5 disc lesion but patient opted for conservative treatment. She was started on celecoxib 200 mg per day and pregabalin 75 mg twice a day and prescribed exercise programme and educated in back care ; she reported decrease in pain with this treatment.

Discussion:

Dystonia:

Dystonia is defined as a movement disorder characterised by sustained or intermittent muscle contractions causing abnormal, often repetitive movements, postures, or both. Dystonic movements are typically patterned, twisting, and may be tremulous. Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation [4]. The presentation is often variable. Our patient had generalised dystonia affecting different parts of the body variably including mild dysarthria, painful contractions of trunkal muscles, foot inversion during walking and sustained contraction of the extensor hallucis longus muscle.

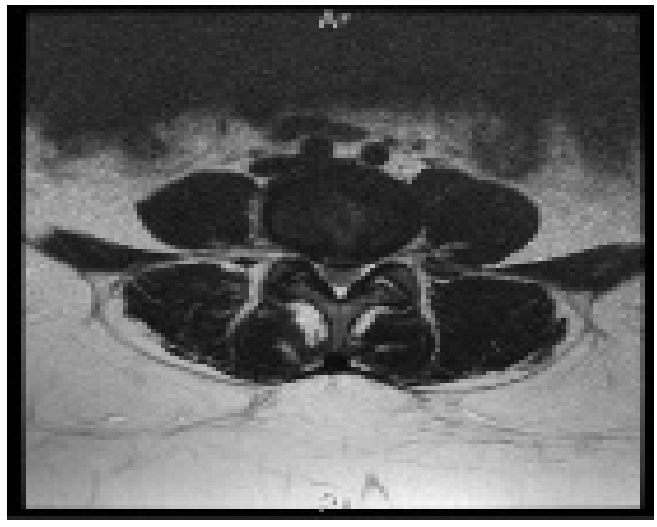

Fig 1: MRI Lumbar Spine, Showing L 4/5 Disc Bulge with Bilateral L5 Root Compression (Sagittal Weighted T2 images)

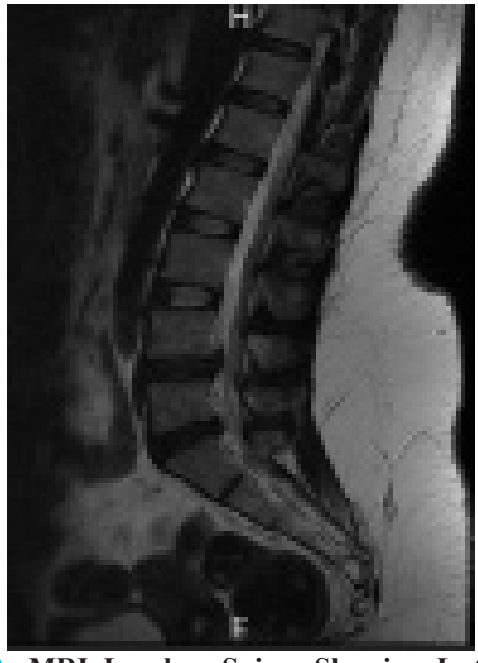

Fig 2: MRI Lumbar Spine, Showing L 4/5 Disc Bulge with Bilateral L5 Root Compression (Axial T2 Weighted Images)

Disabilities due to Hitchhiker's toe:

Hitchhiker's toe is disabling because of pain, shoe difficulties and abnormal posture.Complications include instability and falls. Our patient sufferred from recurrent ankle sprains. The deformity was was so severe that it was resulting in damage to her shoes.

Management of Hitchhiker's toe:

Surgery was the only effective strategy to manage EHL hyperextension before the introduction of botulinum toxin. Recent studies have proved that botulinum toxin

A is effective in the management of Hitchhiker's toe[5]. Yelnik *et al* [6] described a case series of 11 patients with hemiplegia and overactivity of EHL; with post botulinum toxin injection, EHL overactivity disappeared and there was good subjective improvement of pain and shoe difficulties. The authors also described the scale to assess outcome after treatment which we have applied in this report. In our patient with generalised idiopathic dystonia, botulinum toxin A injection to EHL was effective in the management of Hitchhiker toe ; there was decrease in pain and shoe difficulties improvement in the severity of the overactive EHL and improvement of gait.

Weakness following botulinum toxin injection:

Botulinum toxin causes weakness in the muscle by neuromuscular blockade. After injection the toxin diffuses into the muscle and adjacent tissues. Adjacent muscle weakness can occur as side effect if high volumes are used[7]. Generalised weakness due to toxin spreading in blood is very rare. Our patient developed weakness in extensor hallucis longus and extensor digitorum on the left side. Ankle dorsiflexion and plantar flexion were normal. However the history of acute episode of low back pain with radiation to left lower limb, presence of sensory impairment in L5 and MRI finding of L4/5 disc lesion with L5 root compression confirmed that the weakness was the result of the disc lesion and not a side effect of botulinum toxin.

Spinal lesions and dystonia:

There are reports about cervical spine degenerative changes in patients with movement disorders. Hirose and Kadoya[8] proposed that chronic involuntary movements seen in dystonia and athetosis contribute to the early degenerative changes in cervical spine. The mean age in their review of 251 cases was 33 years. Waterston *et al* [9] reported two cases of idiopathic dystonia with cervical spondylotic myelopathy. A search in Pubmed did not find any report on a causal relation between dystonia of trunk muscles and lumbar disc lesion. Our patient was 40 years old with history of dystonic movements since childhood ; the movements predominantly involved the trunk and lower limb muscles. Her back pain had started at the age of 35 years . It can be hypothesised that the repetitive dystonic movements could have been a contributory factor for the early development of lumbar disc lesion with neurological deficit.

Hitchhiker's toe and lumbar disc lesion:

L4/5 disc lesion can present with Hitchhiker's toe. Blunt *et al*[10] reported a patient with dystonic contraction of bilateral great toe extensors and MRI findings of marked lumbar canal stenosis and bilateral L5 and S1 roots compression. The dystonic movements were ameliorated after spinal decompression. They hypothesised that lumbar disc lesion can produce foot dystonia. In our patient the dystonic movements had preceded the low back pain by several decades and the foot dystonia was part of the picture of generalised dystonia.

Conclusions:

Botulinum toxin A injection is effective in the management of dystonic Hitchhiker's toe. When unexplained lower limb weakness occurs in lower limb after botulinum toxin injection in dystonic patient, a careful search for lumbar disc lesion should be made.

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Original Article

Predicting Factors For Development of Heterotopic Ossification in Spinal Cord Injury Patients

Pabitrakumar Sahoo[1], Mamatamanjari Sahu[2], D K Singh[3], S P Das[4]

ABSTRACT

Heterotopic ossification (HO) is a frequent complication in spinal cord injury (SCI) that is often difficult to treat. the incidence ranges from 16-35%. Association of certain complications in spinal cord injury increases the incidence of HO. This is a cross-sectional study conducted at SVNIRTAR, Cuttack, Odisha from January 2009 to December 2014. Out of 132 SCI patients attended to OPD or admitted to SCI ward, HO at different site was diagnosed in 86 patients. A close observation was done on these patients with respect to their associated complications and their relation to development of HO is discussed.

Key wards: **Heterotopic ossification(HO), spinal cord injury (SCI), spasticity, pressure sore.**

Introduction:

Heterotopic ossification (HO) is the formation of true bone in ectopic sites often within the connective tissue of muscle surrounding peripheral joints in patients with neurologic disorders.. It is a frequent complication in spinal cord injury (SCI)patients where the incidence ranges from 16-35%. In non-traumatic myelopathies the incidence of HO seems less compared to traumatic SCI ranging somewhere between 6-15%. HO is less common in children than in adults with an incidence generally reported between 3and10%[1,2].

According to Chalmers *etal*[3] three conditions must be met for the formation of ectopic ossification: the presence of osteogenic percursor cells, an inducing agent, and a permissive environment. Although the precise causal mechanism for HO is still unknown, humoral, neural and local factors probably all play a role in its pathophysiology . There is either a migration of distant mesenchymal cells to the area involved, with subsequent transformation of these cells into osteoblasts, or a transformation of the local mesenchymal cells directly into osteoblasts[4-6]. Whether these cells migrate at random or in response to some chemotactic factor is still not known, but the importance of several factors has been suggested in the transformation of mesenchymal cells into osteoblasts[6].

Microscopic study shows that it has four zones.

1. Innermost zone contains highly active zone with mitotic figures like malignant sacomatous cells.
2. Adjacent zone containing cells less active in appearance, but forming osteoid tissue.
3. Zone of trabecular organisation with osteoblasts and fibrous tissue.
4. Peripheral zone of fibrous tissue.

The purpose of the study is to find out possible associated factors which can be considered as predictors of development of HO in SCI patients. The data are compared with data of published literature.

Materials and Methods:

This cross-sectional study was done during January 2009 to December 2014. All the patients attending OPD as OPD patient and patients admitted to SCI ward of SVNIRTAR, Olatpur, Cuttack were screened for HO. Patients were assessed clinically and radiologically.

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Clinical diagnosis of HO was made in patients presented with unexplained, peri-articular swelling with local warmness, erythema, sometimes accompanied by a low-grade fever and pain in patients with incomplete lesion and sensory sparing, decreased joint range of motion to complete ankylosis of one or more large joints of body. X-ray of the affected joint was done to confirm the diagnosis. Radiological finding varies from an increased density of the peri-articular soft tissues to complete mature extra osseus cortical and trabecular new bone formation. Blood investigations like DC,TLC, ESR, CRP, serum alkaline phosphatase were done to support the diagnosis in radiological inconclusive cases and also to rule out conditions like infection, DVT or impending pressure ulcer.With these features, out of 132 SCI patients attended to OPD or admitted to SCI ward,HO at different site was diagnosed in 86 patients. Hence total no of patients included in the study were 86; 67(78%) were male and 19(22%) were female. Age group range 12-58 years with mean age of 32 years. Data were collected in relation to the following factors-

- Diagnosis of HO from day of injury
- Nature of lesion -- complete / incomplete
- Site of lesion -- cervical / thoracic / lumbar
- Surgery -- non-operated / spinal stabilisation surgery
- Pressure sore -- grading
- Urinary tract infection -- symptomatic/ asymptomatic
- Spasticity -- MAS grading
- Large joints affected

SCI patients with pressure sore of grade III and grade IV were included for data collection. As per European Urological Association criteria of diagnosing UTI, SCI patients presenting with fever, suprapubic pain, turbid urine, haematuria and leucocytes in urine of >40 leucocytes/mm3were included as symptomatic UTI.

Modified Ashworth scale was used for spasticity grading. Pressure sores present over various pressure bearing areas like sacrum, trochanter and ischium were graded by standard grading fixed by National Pressure Ulcer Advisory Panel(NPUAP).

Results:

Average duration of diagnosis of HO from date of injury is 5 and 1/2 months ranging from 2 months to 9 months. On clinical evaluation 54 cases(62.7%) showed complete injury and 32 cases(37.3%) showed features of incomplete injury (Fig 1). Radiological site of lesions are- cervical-31(36%); thoracic 49 (60%) and lumbar 6 (7%) cases (Fig 2). 34 cases (39.5%) had undergone surgery for spinal fixation, 52 cases (60.5%) had not undergone any form of spinal fixation. Pressure sore of grades III and IV were present in 52 cases (60.5%). Eighty cases(93%) had indwelling catheter and 21cases (24%) of them had features of urinary tract infection. All the cases with HO presented with spasticity of different grade as per modified Ashoworth scale. Moderate to severe spasticity of MAS 2, MAS 3grade was marked in 56 cases (65%). HO was found around most of the large joints like hip in 48cases (56%) (Fig 3), knee 25cases (29%)(Fig 4) and around elbow in 13 cases(15%) (Fig 5).

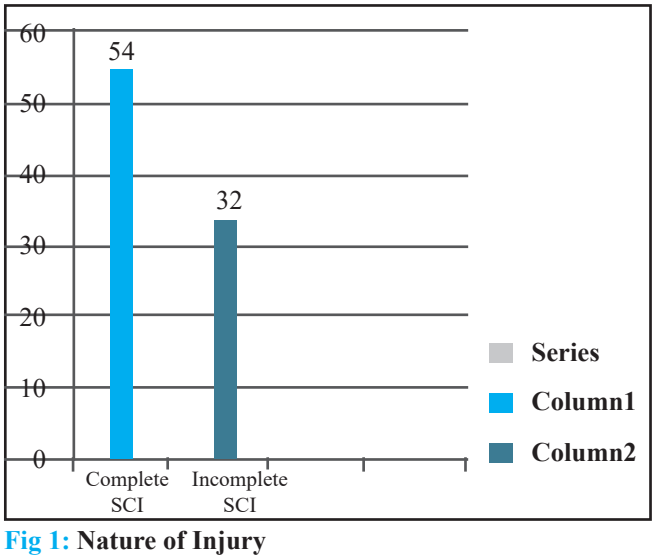

Fig 1: **Nature of Injury**

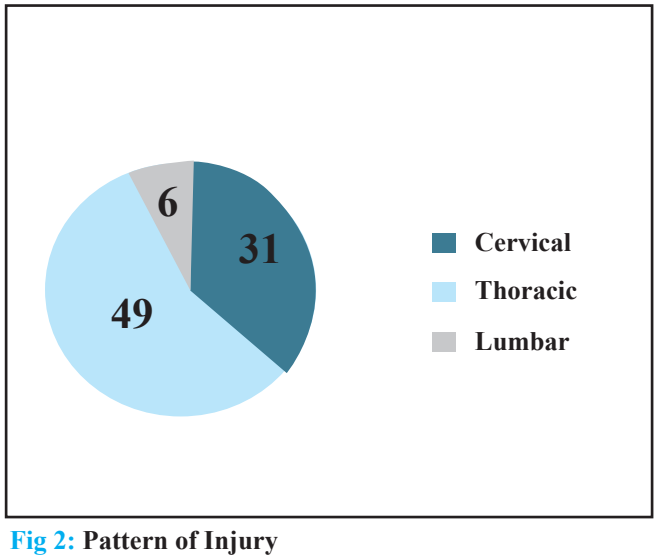

Fig 2: **Pattern of Injury**

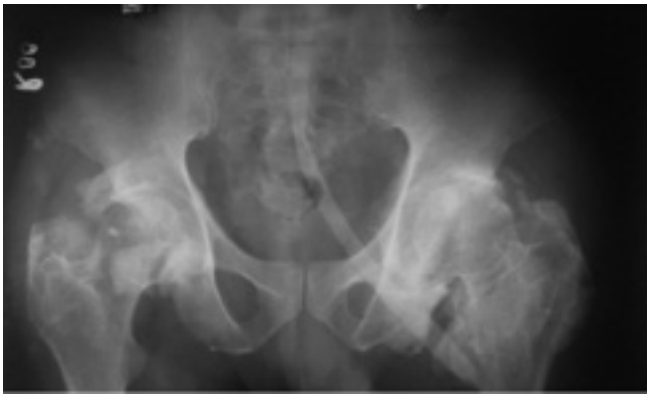

Fig 3: Heterotopic Ossification Around Both Hip Joint

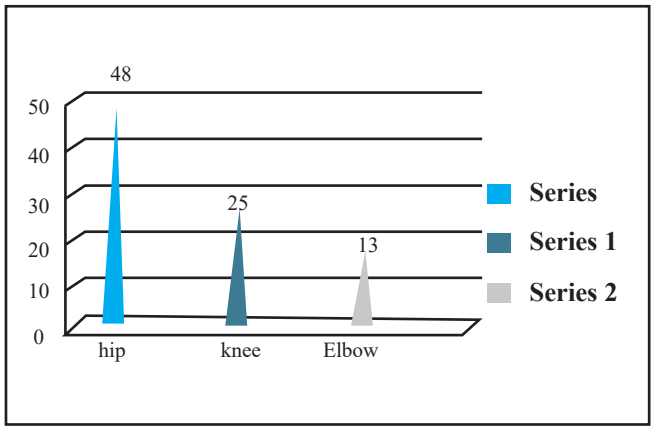

Fig 5: Major Joints of Body Involved with Heterotopic Ossification

Discussion:

The clinical presentation of HO in SCI patients ranges from an incidental finding on x-ray to severe restriction of joint range of motion. Peri-articular swelling with red and induration of the part may be noticed due to interstitial oedema of the soft tissues in acute cases associated with low grade fever with local rise of temperature. In patients with sensory sparing, the first symptom may be pain in the affected area. Although HO may develop even several years after SCI, it is generally diagnosed between 1 and 6 months post-injury with a peak incidence at 2 months[7-12]. HO always occurs below the level of the SCI, most commonly at the hip (70-97%)[13-16], 56% of cases of HO was seen around hip in this study. Although Catz *etal* [17]. did not find a relationship between radiologically diagnosed HO and the severity of the motor deficits. Several authors have reported that complete transverse SCI is more commonly associated with HO than incomplete SCI. This study shows 54 cases (62.7%) of HO in complete spinal injury patients. SCI patients with lumbosacral or cauda equine lesion

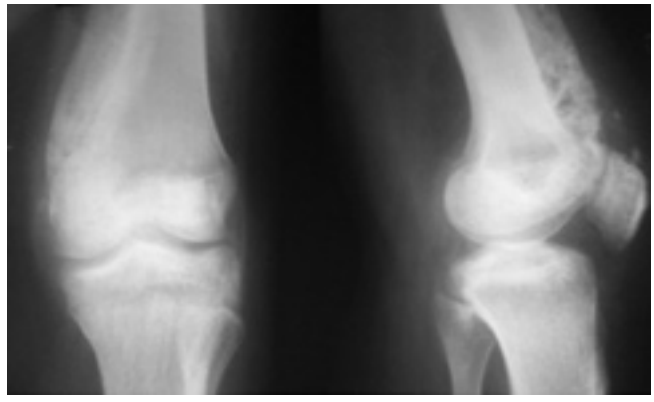

Fig 4: Heterotopic Ossification Around Knee Joint

less frequently reported with HO[18] which is comparable with this study of 6 cases(7%). An area of soft tissue damage due to pressure ulceration with subsequent oedema may predispose to the development of HO [19]; 60.5% of cases presenting with HO had pressure ulcer of grade III or grade IV. An infected urinary tract could serve as a source of antigenic material precipitating an immune response that triggers subsequent HO[20]. However this study shows only 24% cases HO had features of UTI although 97% cases had bladder with indwelling catheter. Controversy also exists regarding the possible association between HO and spasticity. In some studies HO is more commonly seen in SCI patients with spasticity and more extensively in those with severe spasticity[21-23]which is seen in 65% cases in this study.

Conclusions:

Since the pathophysiology of HO is poorly understood, the only preventive treatment possible is the early identification and adequate treatment of the possible risk factors. With adequate nutrition and nursing management, the incidence of urinary tract infections, decubitus ulcers may be reduced, and thereby, the risk of developing HO. Although in the early literature, aggressive passive physiotherapy has been recommended to improve joint mobility and to counteract ankylosis in the case of contractures, it is now generally accepted that SCI patients profit from early, regular, and cautious joint mobilisation. More rigorous exercises with the risk of (micro) trauma to the peri-articular tissues better to be avoided. When gentle passive movements of the large peripheral joints are started and maintained from the day of the injury, the joint capsules are kept as supple as possible, muscles will not easily shorten and contractures will not readily develop, so that HO may be prevented.

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Case Report

A Rare Case of Bilateral Spontaneous Intracerebral Haemorrhage Presenting With Left Hemiplegia: A Case Report

Annada Sankar Mohes[1], L Dorendrojit Singh [2], Aten Jongky[3], Th Bidyarani[4]
L Nilachandra Singh [5], AK Joy Singh [6]

Abstract

Haemorrhage is responsible for around 11% of stroke syndrome. Haemorrhage usually occurs at a single site. However, it can be at multiple sites in some specific conditions i.e. coagulopathy, vascular malformation, malignancy etc. A 56-year-old male with left sided hemiplegia was admitted in the rehabilitation ward of RIMS, Imphal. He was hypertensive and was on irregular medication for that. He was also an alcoholic and chronic smoker for last 20 years. Patient was conscious and clinical examination revealed left 7th and 12th cranial nerve involvement with left hemiplegia. Non-contrast CT scan of brain revealed right thalamus and left basal ganglia haemorrhages. Thorough history and investigations did not reveal any aetiology for bilateral haemorrhage. Patient was treated with conservative management and improvement was noticed in serial follow-ups. There are very few case reports about bilateral spontaneous intracerebral haemorrhage associated with other diseases like migraine, Japanese encephalitis etc. Cause of bilateral haemorrhage in our case is doubtful.

Key words: **Intracrebral haemorrhage, stroke syndrome, cranial nerve.**

Introduction:

Stroke is one of the commonly encountered diseases in rehabilitation ward. Prevalence of stroke is increasing day by day due to rise in elderly population. Infarction and haemorrhage due to vascular cause are responsible for stroke syndrome. Out of these two, spontaneous haemorrhage is responsible for around 11% cases[1]. Putamen is the most common site for spontaneous intracerebral haemorrhage followed by thalamus[1]. Among the haemorrhagic strokes, 10% spontaneous haemorrhage occurs in the cerebellum[1].

Bilateral spontaneous intracerebral haemorrhage is rare in hypertensive patients but can occur in association with other aetiologies. Prognoses of such cases are also poor.[2] However, in our case, no specific aetiology was identified and improvement of the condition was noticed over time.

Case Report:

A 56-year-old male was admitted in the rehabilitation ward with complaint of sudden onset of weakness of left upper and lower limbs. Patient was a known hypertensive patient for last 10 years and was on irregular medication. He was also an alcoholic and chronic smoker for last 20 years. There was no history of any chronic medication, bleeding disorder or any other significant health problem in the past. Clinical examination revealed left 7th and 12th cranial nerves involvement with normal higher mental function and speech. Left upper limb was found to be flaccid. Left lower limb also shows poor motor control with grade 2 motor power in hip flexors according to Medical Research Council scale. Deep tendon reflexes were found within normal limits along with extensor plantar response on affected side. Ankle clonus was present on the left side and sensory response

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was intact on both sides. Blood pressure was 200/120 mm Hg on admission but became normal following medication within few days.

All routine investigations were found to be normal except low HDL level in lipid profile picture. Non-contrast CT scan of brain showed right thalamus and left basal ganglia haemorrhage (Fig1). There was no past history of any abnormal health condition. We investigated the coagulation profile, which was also found to be within normal limits.

Patient was treated conservatively and improvement was noticed during serial follow-ups.

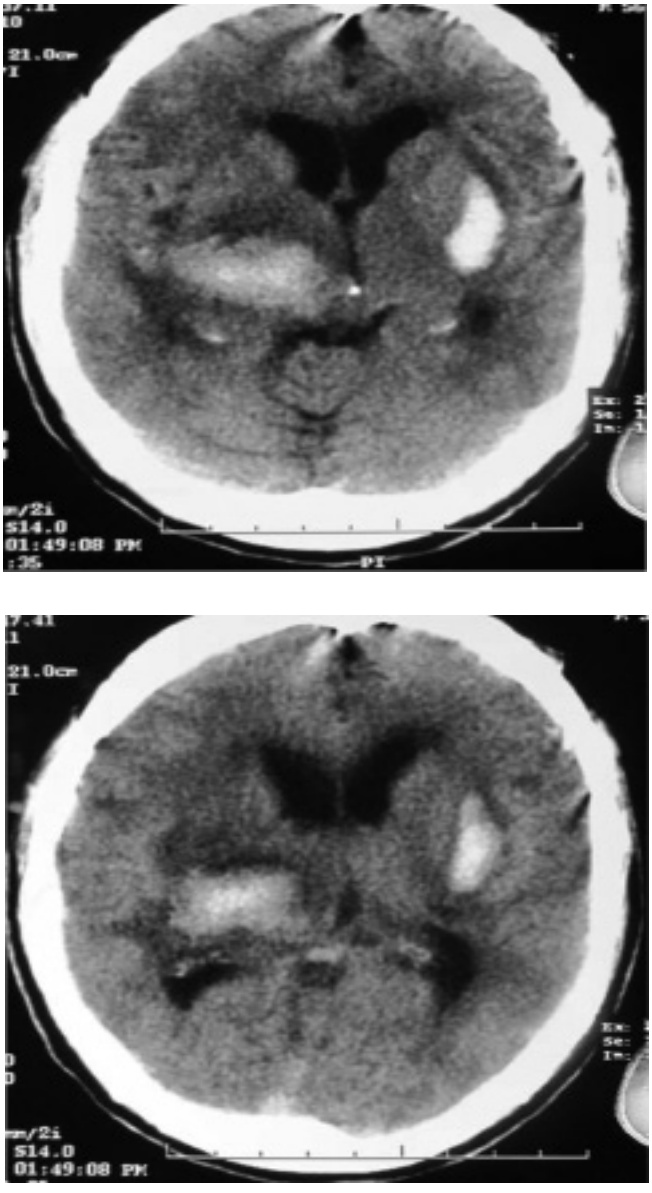

Fig 1: NCCT Brain Shows Bilateral Cerebral Haemorrhage

Discussion:

Stroke is defined as sudden onset of persisting focal neurological deficit of vascular origin[1]. Other causes of focal neurological deficit i.e. brain tumour, abscess, trauma, seizure, etc, are not included in stroke and they form the differential diagnosis of stroke.[1] Risk factors of stroke can be divided into modifiable and non-modifiable ones[1]. Identification of modifiable risk factors are very important as prevention of these factors can prevent stroke and its recurrence. The common modifiable risk factors are hypertension, heart diseases (valvular, arrhythmia etc), smoking, diabetes mellitus, hyperfibrinogenaemia, hyperlipidaemia etc[3]. Among these risk factors, hypertension is the most common aetiology for spontaneous intracerebral haemorrhage[4]. Simultaneous intracerebral haemorrhage can occur in opposite side of brain but bilateral spontaneous intracerebral haemorrhage is a rare phenomenon and few case reports mentioned some association other than hypertension. These are coagulopathy, vasculitis, infection, methanol intoxication, diabetic keto-acidosis, migraine etc[3]. Prognosis of such cases were also found bad even in smaller size haemorrhages[2]. Actual pathophysiology leading to bilateral spontaneous intracerebral haemorrhage is not well known. There are two opinions, one is rupture of microaneurysms bilaterally, formed as a result of chronic hypertension and the second one is simultaneous haemorrhage to opposite brain following first haemorrhage[4]. In our case, we could not find any association. Patient was hypertensive for 10 years and on irregular antihypertensive medication. Therefore, it can be because of rupture microaneurysm formed because of chronic hypertension.

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Pictorial CME

Normal Pressure Hydrocephalus Presented with Confusion and Retropulsion

Pramanik R

An elderly lady with multiple comorbiditis like hypertension, diabetes and dyslipidemia with past history of stroke presented to us in our OPD with gradually progressive confusion, minimal memory disturbances and loss of balance over a period of 6 months. Her son complained about her slowness of movement, lack of initiation and gradual stooping posture. On thorough clinical examination in indoor we didn't find any significant rigidity or tremor in her extremity, her postural blood pressure was absolutely fine but she had a definite retropulsion along with depression. On close questioning she gave a history of increased frequency and few episodes of incontinence in last 15 days.

Considering the above clinical picture we sent the urine and blood samples and found out that she had leucocytosis with increased inflammatory markers, and her urine culture revealed E.coli infection with a colony count of 107. The patient was started on antibiotics and the patient's confusion decreased (MMSE from 20 to 25), but the patient still had the presenting symptoms. This prompted us to do a MRI brain to rule out dementia due to multi-infarct state. Mean while the patients gait pattern and walking showed signs of improvement after non pharmacological management like balance and gait training, strengthening of the lower limbs.

MRI (fig 1 & 2) showed no definite hydrocephalus but Evans index of 32 (radiological fulfillment of NPH), a CSF tap was scheduled to look for opening pressure and it was 270 mm of water, so a therapeutic drainage of 30 ml was done. Following this the patients gait pattern as well as walking speed improved (10 meter walking time from 15 s to 12 s) and MMSE improved to 28 from 25.

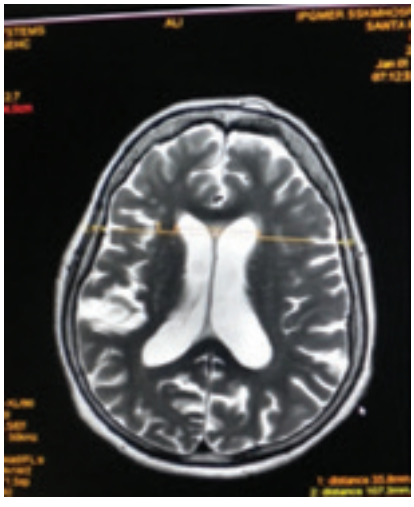

Fig 1

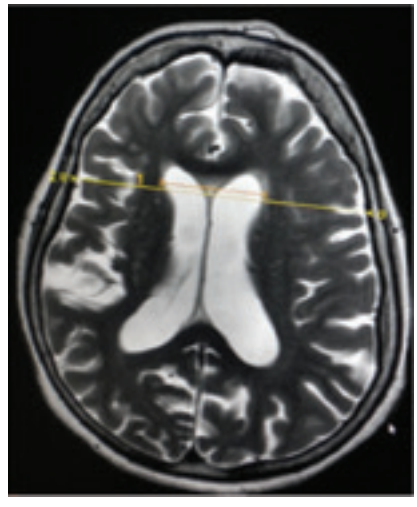

Fig 2

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Cite as:
Pramanik R. Missed Opportunity For Thrombolysis In A Patient of Massive MCA Territory Stroke. IJPMR, December 2015; Vol 26(4) :122

PG Forum

REHAB CHALLENGE

An 16 year old girl from poor socioeconomic status studying in class x presented in PMR OPD with sudden onset severe weakness of right upper limb (Fig 1) preceded by severe indirect injury around her right shoulder 2 ½ years back. Initially she was treated with hyper-abduction splint, exercise therapy and electrical stimulation for few months. But there is very poor clinical response from the above regimen. Electrodiagnostic study of her both upper limb was also corroborative with her clinical status.

Later on a tendon transfer surgery (Trapezius to Supraspinatus) (Fig 2) was done aiming independent shoulder abduction, unfortunately she didn't receive a proper post operative rehabilitation regimen for independent functioning of right upper limb. After that she was treated intermittently by different specialist in several hospitals without any positive response.

When we examined the patient (fig 3), we found a muscle power of 1/5 in Supraspinatus and deltoid, 3/5 trapezius, 0/5 in biceps , 1/5 in triceps, 3/5 in wrist extensors and 3/5 in intrinsic muscles of hand. This young girl is very keen to perform independent ADL by her right upper limb.

Please opine regarding further rehabilitation plan for this patient.

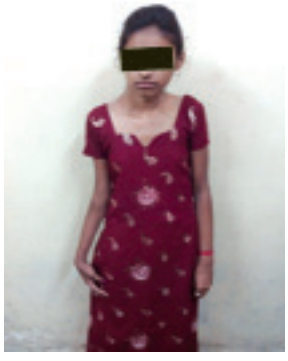


Fig 1

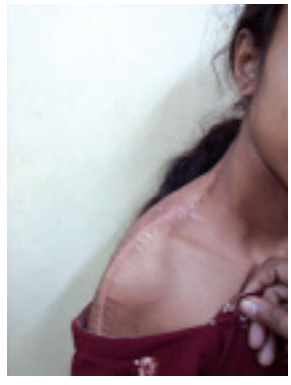
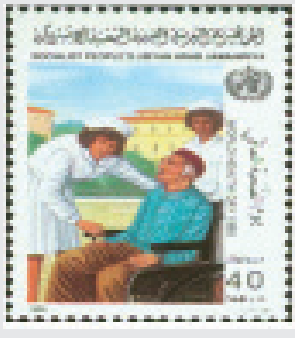


Fig 2

Fig 3

Medical Philately

Libya 1985 World Health Day SG1668 Wheelchair / Elderly



Country	Libya
Date	1985
Disability	Wheelchair
Meta	stamp, disability, outside centre, Libya, 1985, World, Health, Day, SG1668, Wheelchair, Elderly
Number	sg1668

Country	Pakistan
Date	2003-12-03
Disability	Crutches
Meta	philately, stamps, digital disability, theme, imagery of disability on postage stamps, outside centre, crutches, integration, friends, social integration
Theme	Integration International Year of Disabled People
Size:	29 x 34 mm
Face value:	2 RP - Pakistani rupee

PG Forum

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When we examined the patient (fig 3), we found a muscle power of 1/5 in Supraspinatus and deltoid, 3/5 trapezius, 0/5 in biceps , 1/5 in triceps, 3/5 in wrist extensors and 3/5 in intrinsic muscles of hand. This young girl is very keen to perform independent ADL by her right upper limb.

Please opine regarding further rehabilitation plan for this patient.

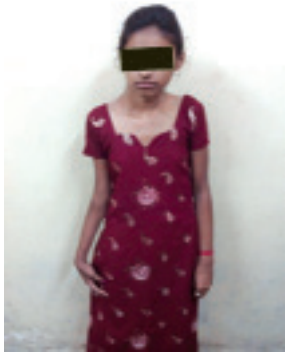


Fig 1

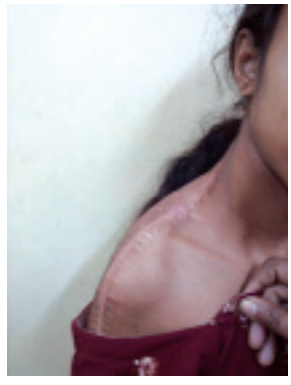
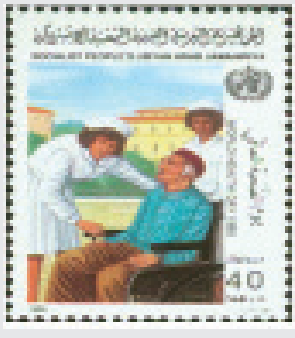


Fig 2

Fig 3

Medical Philately

Libya 1985 World Health Day SG1668 Wheelchair / Elderly



Country	Libya
Date	1985
Disability	Wheelchair
Meta	stamp, disability, outside centre, Libya, 1985, World, Health, Day, SG1668, Wheelchair, Elderly
Number	sg1668

Country	Pakistan
Date	2003-12-03
Disability	Crutches
Meta	philately, stamps, digital disability, theme, imagery of disability on postage stamps, outside centre, crutches, integration, friends, social integration
Theme	Integration International Year of Disabled People
Size:	29 x 34 mm
Face value:	2 RP - Pakistani rupee

PG Forum

REHAB QUIZ

1. **The highest complete SCI level that can live independently without the aid of an attendant is a:**
   a. T10 complete tetraplegia
   b. C6 complete tetraplegia
   c. C5 complete tetraplegia
   d. C7 complete tetraplegia

2. **Quadriceps weakness suddenly improves with exercise and Guanidine in**
   a. Polymyositis
   b. Dermatomyositis
   c. Myasthenia gravis
   d. Lambert-Eaton syndrome

3. **The trim lines of a ground reaction ankle foot Orthosis should be anterior to the malleoli to serve as a:**
   a. Dorsiflexion assist
   b. Plantar flexion assist
   c. Dorsiflexion stop
   d. Plantar flexion stop

4. **Which medication is preferred for spasticity of cerebral origin?**
   a. Flexeril
   b. Baclofen
   c. Dantrolene
   d. Diazepam

5. **In an acute shoulder dislocation, how long should the shoulder be immobilized after it has been reduced?**
   a. 6-8 weeks
   b. 4-6 weeks
   c. 1-3 weeks
   d. No immobilization is needed

6. **Which anatomical structure is most likely damaged in a stroke patient who has aphasia, consisting of fluent speech, impaired comprehension and preserved repetition of words?**
   a. Parietal operculum
   b. post superior temporal gyrus
   c. temporal occipital cortex
   d. post inferior frontal lobe

7. **In thoracic outlet syndrome, compression most commonly occurs at the:**
   a. Axilla
   b. Interscalene triangle
   c. Subcorocoid space
   d. Costoclavicular triangle

8. **Valsalva manoeuvre in rehabilitation regimen is a major concern in**
   a. Ischemic Cardiomyopathy
   b. Hypertrophic Cardiomyopathy
   c. Dilated Cardiomyopathy
   d. Duchene's Cardiomyopathy

9. **Which compartment is most commonly affected in exertional compartment syndrome?**
   a. Lateral b. Medial
   c. Anterior d. Posterior

10. **What is the MOST important factor in decreasing the vertical loading of the lumbar spine?**
   a. Application of a three-point pressure system for vertical stabilization A.
   b. Enhancement of the abdominal hydro pneumatic mechanism
   c. Strengthening of the glutei and abdominal musculature
   d. Immobilization of the thoracolumbar spine

ANSWERS

Answer of September 2015

1B, 2C, 3B, 4D, 5C, 6B, 7B, 8C, 9A, 10B

Case Report

Stroke Presenting as an Isolated Hand Palsy

G. Sonachand Sharma[1], Y. Nandabir Singh[2], Alex T Touthang[3], Tamphleima Kh[4]

Abstract

Isolated hand palsy also known as 'pseudoperipheral palsy' is a rare presentation of ischaemic stroke, often mistaken for peripheral nerve lesion. Here, we report a 13 years old young girl presented with sudden onset right hand palsy without any typical features of either upper motor lesion or lower motor lesion. Ischaemic stroke caused by embolic infarct of left precentral gyrus was the possible cause for her. She was managed with physiotherapy interventions including electrical stimulation, strengthening, grip exercise etc. Such an isolated hand palsy resulting from stroke is a rarely reported entity.

Key words: **Isolated hand palsy, ischaemic stroke, pseudoperipheral palsy.**

Case Report:

A 13 years old girl presented with sudden onset of complete paralysis of right hand (Fig 1) without any sensory disturbances for a period of 10 days. The patient noticed the weakness of right side of the body when she wake up early in the morning that was associated with headache, paresthesia and weakness in speech. However when she reported to us, she had only predoninant weakness of right side of the hand disturbing in her activities of daily livings. There was no history of trauma or any loss of consciousness. Bladder and bowel control and higher mental functions including speech were normal at the time of admission. She had tuberculous meningitis with parasitic neck infections in 2008 and it was treated with medications. The patient was consulted first to the neurologist where she was treated in the line of cerebro vascular accident. Latter, she reported to us for further management.

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Clinical examination of right hand revealed muscles weakness with MRC (medical research council) scale 0/5 in the muscles beyond the wrist joint without any wasting of muscle. But, proximal forearm and arm muscles, sensation, muscle tone, and tendon reflexes were unaffected. However, Hoffman sign was positive on the affected hand. Further clinical examination including function of language, cranial nerves, and plantar reflex, did not show any abnormalities.

Routine baseline investigations test were within normal. Chest x-ray and neck x-ray did not show any abnormal features. Electromyelogram showed normal nerve conduction velocity and normal late responses. Vaculities and antiphospholipid workup were negative. EEC and echocardiogram were also normal. Retro-viral test was also non-reactive. NCCT scan of brain revealed small left temporoparietal infarct (Fig 2). Diffusion-weighted MRI brain and angiogram could not be done due to financial constraints. Psychiatry consultation did not establish any possible disease.

Proper counselleing regarding disease condition was done to patients as well as care giver. For paralysed hand, we advised electrical stimulation followed by active assistive strengthening exercise gradually shifted to active resistive and grip strengthening exercises. In order to prevent complications, we also advised range of motion exercise of hand joints and stretching exercise of finger flexors. After a month, the patient reported with improved right hand muscles power with MRC scale of 4/5 and nearly normal in her activities of daily living.

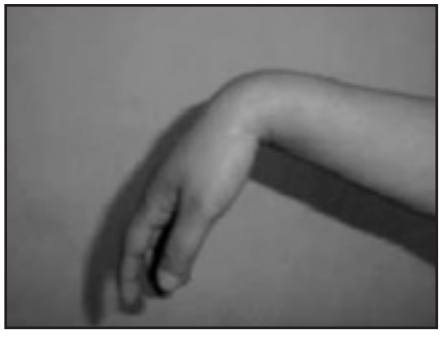

Fig1: *Isolated Hand Palsy*

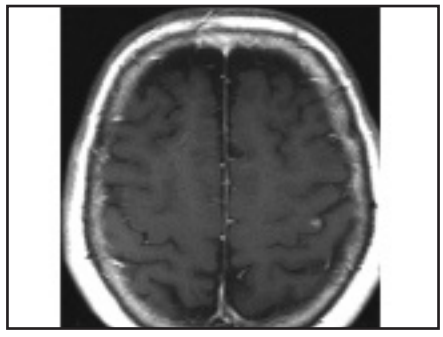

Fig 2: *Ischaemic Infarct*

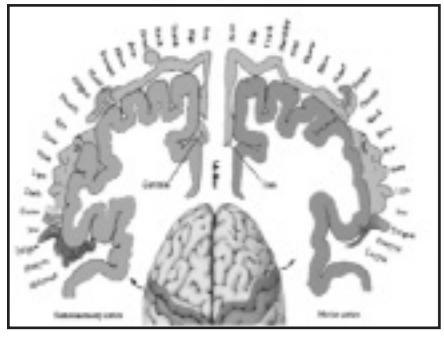

Fig 3: *Motor Homunculus*

Discussion:

Motor homunculus (Fig 3) is the pictorial representation of body's anatomical part in the bran. It is located in primary motor cortex area of cerebral cortex in precentral gyrus. In the motor homunculus, the hand occupies big area. This hand knob area is supplied by small cortical branch from superior division of middle cerebral artery. Ischaemic infarct in this hand knob area may lead to hand palsy.

Isolated hand palsy also known as pseudoperipheral palsy, caused by cerebral lesions is very rare and often mistaken for peripheral lesion[1,2]. The term "fractional arm weakness" has also been applied when weakness of the hand differs from that of the proximal joints[3]. Isolated hand palsy has been reported as a result of, 1) embolic stroke involving the hand knob area, 2) large atherosclerotic infarct of vascular border zones area of cerebral cortex, 3) subcortical lacunar infarct, and 4) rarely due to inferior parietal lobe infarctions resulting from severe carotid stenosis or dissection[3,4].

Celebisoy *et al*[5] reported eight patients presenting with isolated hand palsy due to discrete cortical infarction in the precentral gyrus.

In the present case report, we assumed the involvement of hand knob area in motor homunculus region in the left precentral gyrus by embolic infarct was the possible cause for the isolated hand paralysis. The possibility of peripheral nerve lesion was excluded by clinical features and normal EMG findings. Psychiatric problem was also excluded. Even though stroke in young age is rare, source of emboli in our case may be due to either tubercular meningitis or parasitic neck infection which the patient had in the past years.

So, it is important to be aware of the broad differential diagnosis of isolated hand weakness and the ischaemic stroke should also be included as a differential diagnosis for uncommon cause of isolated hand palsy.

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Original Article

Ultrasound: A Screening Tool for Carpal Tunnel Syndrome

Bobeena Rachel Chandy[1], Betty M.[2], Henry Prakash Magimairaj[3], Binu P. Thomas[4], George Tharion[5]

Abstract

Background: Electrodiagnostic test is considered as the gold standard for diagnosis of carpal tunnel syndrome (CTS). Ultrasonography provides a simple non-invasive means of visualising peripheral nerve pathology.

Objective: The objective of the study was to assess the role of ultrasonography in CTS and its correlation with the present day gold standard of nerve conduction studies (NCS).

Materials and Methods: A prospective cohort size of 100 subjects was calculated based on a hypothesized sensitivity of 90% and a confidence interval of 85-95%. All 100 subjects, 64 controls and 36 patients underwent nerve conduction studies and USG. Transverse images of the median nerve were obtained at three levels: proximal to the carpal tunnel inlet, at the carpal tunnel inlet and at the carpal tunnel outlet. The flattening ratio was also assessed at the tunnel inlet and outlet. Statistical analysis was done to corelate the ultrasound findings at each level with nerve conduction studies and calculation of the positive and negative predictive values. The cut offs of the cross-sectional areas of the median nerve at the three anatomical levels on ultrasonography were taken at the best sensitivity and specificity according to the ROC curve.

Results: We found that at any one anatomical level, the sensitivity of ultrasound to detect carpal tunnel syndrome by increase in the cross-sectional area of median nerve as compared to the nerve conduction studies is 90%.

Conclusions: At 45% specificity, ultrasonography could be used as a non-invasive and easily available screening tool in carpal tunnel syndrome. Also, the best level to look for nerve compression is at the level of the carpal tunnel inlet.

Key words: **Carpal tunnel syndrone, ultrasound, median nerve.**

Introduction:

Carpal tunnel syndrome (CTS) is a common upper limb entrapment neuropathy caused by the compression of the median nerve in the wrist[1]. CTS became widely known among the general public in the 1990s because of the rapid expansion of office jobs and increased use of computers. CTS was found to have an estimated lifetime risk of 10% and its prevalence being 5% in the general population with a female preponderance ranging from 3:1 to 23:1[2]. Diagnosis of this condition is mainly based on the history of symptoms (tingling, numbness, pain and burning sensation in the hand), provocative factors (repetitive movement of the wrist, sleep), mitigating factors (shaking the hand, changes in hand posture) followed by electrodiagnostic tests which are considered as the gold standard.

Diagnostic ultrasonography is non-invasive, there is no radiation, its readiness of use, cost-effectiveness and ability to make dynamic examinations possible, makes it a popular investigation tool for musculoskeletal disorders and pain management interventions. Peripheral nerve ultrasonography is emerging as a promising diagnostic tool for entrapment neuropathies particularly CTS, by demonstrating enlargement of the nerve, bowing of the flexor retinaculum, swelling ratio, and increase in the cross-sectional area of the media nerve in the carpal tunnel [3-18] as it provides a simple non-invasive means of visualising nerve pathology.

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Most of these studies, however, have not demonstrated the true diagnostic use of US as the NCS results have been used as reference standards. This study was done to see if ultrasonography can be used as a screening tool for CTS based on the sensitivity and specificity, and also compares the US and NCS for CTS. This study is aimed to find the value of sonography for CTS diagnosis so that it can be used especially in medical set-ups where tests like electrodiagnostic studies and MRI are not easily available.

Materials and Methods:

The study was conducted in the Department of Physical Medicine and Rehabilitation, in a tertiary care hospital in south India. The participants were patients with symptoms of CTS, referred from the out-patient services of the departments of Hand and Leprosy Reconstructive Surgery (HLRS), Orthopaedics and PMR for nerve conduction studies (NCS).

The total sample size of 100 subjects was required to find a hypothesised sensitivity of 90% with a 95% confidence interval of 85-95%. Of this number, 36 were in the patient group and 64 in the control group.

The study was approved by the Institution Review Board of the Institute and an informed consent was obtained from all the participants.

Control Group:

Normative data was collected from 64 age-group matched subjects recruited from the hospital staff or relatives accompanying the patients, who did not have any signs or symptoms of CTS (33 males and 31 females). They were subjected to history and neurological examination to rule out any abnormality. NCS and ultrasonography of both wrists were done for all subjects included in the control group.

Patient Group:

Thirty-six patients were included in the study. The patient group were selected based on the diagnostic criteria put forward by the American Academy of Neurology (1993)[19] – paresthesia; pain; swelling; weakness or clumsiness of hand provoked or worsened by sleep; sustained hand or arm position; repetitive action of the hand or wrist that is mitigated by changing posture or by shaking of the hand; sensory deficit or atrophy of the median nerve innervated thenar muscle; symptoms elicited by the Phalen's test performed on each patient.

A detailed history was taken using the modified Boston carpal tunnel questionnaire, which assessed the symptoms (pain, paresthesia, numbness, weakness and nocturnal symptoms) and functional status (writing, buttoning, holding, gripping, opening jars, carrying grocery bags, household chores, bathing and dressing). This questionnaire was modified to be used in the Indian population by virtue of being translated into Indian regional languages, utilising a translated visual analogue scale and assessing functions (eg, activities like buttons/ hooks/sari pleats) relevant to our population[20].

A clinical examination was done for any sensory deficits in the median innervated area in the hand (categorised as impaired sensations/ normal sensations) and for motor weakness of the abductor pollicis brevis (categorised as weakness present or normal). Tinel's and Phalen's tests known to be highly sensitive (0.97 and 0.92) and specific (0.91 and 0.88) for diagnosis of CTS, was also performed for each subject [21]. The radiologist was blinded to the clinical findings.

Thirteen out of the 36 patients were found to have predominant motor symptoms and 23 people were found to have mainly sensory impairment in the hand in the median innervated areas, namely the thumb, index, middle and radial half of the ring finger. Laboratory investigations (blood sugars and thyroid functions) were also done to rule out any other cause for CTS. Informed consent was taken from all the patients.

Electrodiagnostic evaluation:

Electrodiagnostic studies were performed for all subjects included in the study according to the protocol put forth by the American Association of Electrodiagnostic Medicine recommendations [19,22,23] using Medelec Synergy (VIAsys Healthcare EMG and EP systems, UK Ltd, software version 11).

The nerve conduction study was carried out by the same physician, in the elecrodiagnostic lab with specified ambient room temperature of 32 degrees. Standard tests for median sensory and motor conductions, included recording of distal latency, conduction velocity across the wrist and amplitude.

The criteria for diagnosis of CTS were [19, 23-25] :

-Distal sensory latency recorded from the index finger (antidromic stimulation) using ring electrodes is >3.3 ms.

- Distal motor latency of median nerve recorded from the abductor pollicis brevis using disc electrodes, with stimulation 3cm proximal to the distal crease of the wrist > 4.4 ms.

When standard tests mentioned above yielded normal results, a comparative median/ ulnar studies were done.

- Difference between distal motor latency of median and ulnar nerves > 1.1ms
- Difference between distal sensory latency of median and ulnar nerves > 0.2ms[23].

F wave was done for all patients.

In accordance to with the results of the electrodiagnostic studies, the hands were categorised into 3 groups:

1. Mild to moderate CTS – characterised by slowing of the median sensory distal latency with normal distal motor latency or abnormal distal motor latency with or without delay in conduction velocity and diminished amplitudes.
2. Severe CTS – absence of a median sensory response and prolonged distal motor latency with delayed conduction velocity and diminished amplitude or absence of CMAPs.
3. Normal – motor and sensory distal latency, conduction velocity and amplitude within normal limits.

Sonography:

All 100 subjects underwent high-resolution ultrasound performed by two radiologists who were experienced in the field of musculoskeletal sonography using Seimens Antares Ultrasonography machine, with a 7-13 MHz linear array transducer. The radiologist was blinded to the clinical findings and electrodiagnostic results. The sonographic examination was done within 3 days of the electrodiagnostic study. The examination was performed with the patient seated in a comfortable position facing the radiologist with the forearm resting on the table in supination with the wrist in the neutral position and fingers semi-flexed. Transverse images of the median nerve were obtained at three levels: Immediately proximal to the carpal tunnel inlet (distal radio-ulnar joint level), at the carpal tunnel inlet (level of scaphoid and pisiform) and at the carpal tunnel outlet (trapezium and hook of hammate level). The cross-sectional area was measured by tracing it with an electronic calliper around the margin of the median nerve (Fig1). The flattening ratio, defined as the ratio of the major axis of the median nerve to the minor axis, was also assessed at the tunnel inlet and outlet (Fig 2). Of the 100 subjects included in the study (36 patients with CTS and 64 normal subjects), 18 were found to have unilateral bifid median nerve (Fig 3). Among the patient group, there were 8 people with this variant but in the asymptomatic hand. Among the normal subjects, 10 individuals had the variant, hence the wrist with the variant nerve was excluded from the study and the normal wrist was included.

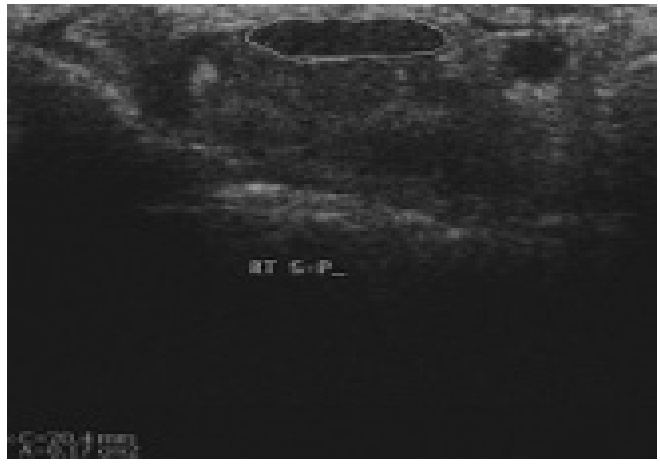

Fig 1: Cross-sectional Area of the Median Nerve Measured by an Electronic Calliper

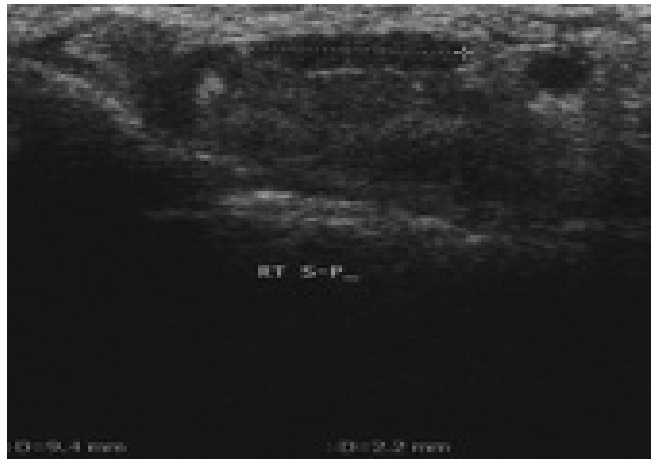

Fig 2: Flattening Ratio (Ratio of Major Axis of Median Nerve to the Minor Axis).

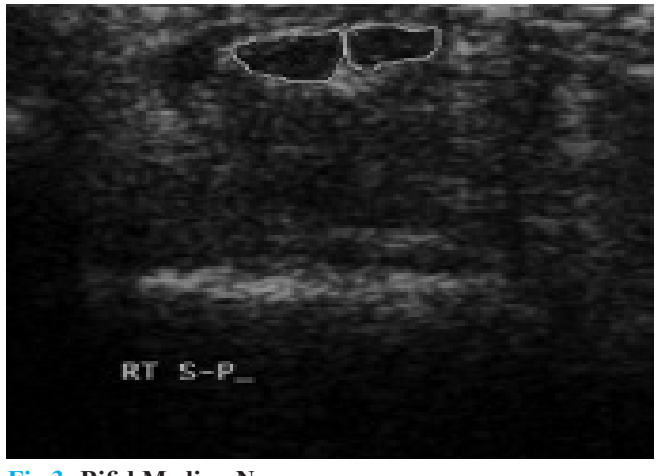

Fig 3: Bifid Median Nerve.

Statistical analysis:

The statistical analysis was done using the STATA 8.0. The cut off values of the cross-sectional area for each of the three levels (proximal to inlet, at the inlet and at the outlet) were calculated according to the receiver operating characteristic (ROC) curves for each level (Figs 4-6). The co-relation of the positive ultrasound findings at each level and nerve conduction studies was done along with calculation of the positive and negative predictive values. The mean flattening ratio for cases, controls and the combined group were also calculated.

Results:

Among the 36 patients recruited for the study 10 were males and 26 were female and among the control population of 64 subjects, there were 33 males and 31 females. The age distribution in the patient population was between 18 and 56 years with a mean of 38.33. Among the control group the age distribution was 18-58 years with a mean of 29.89.

A total of 177 wrists were studied, 59 from the patient group and 118 of the control group. In the patient group, 23 subjects were found to have bilateral CTS and 13 were found to have unilateral disease (11 right sided and 2 left sided).

Among the patient group, 29 were classified under group 1 (mild to moderate CTS), 7 individuals were classified under group 2 (severe CTS), and 64 in group 3 (normal findings. Eighteen subjects among the cases as well as the control group were found to have a bifid median nerve, which has been described in literature as a normal variant [26].

Receiver operating characteristic (ROC curve) analysis was done for the cross sectional areas for the 3 levels of measurement for the best cut-off values.

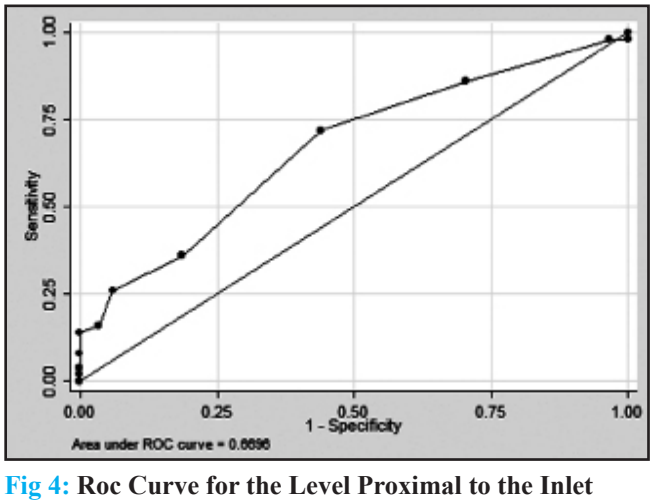

Fig 4: **Roc Curve for the Level Proximal to the Inlet**

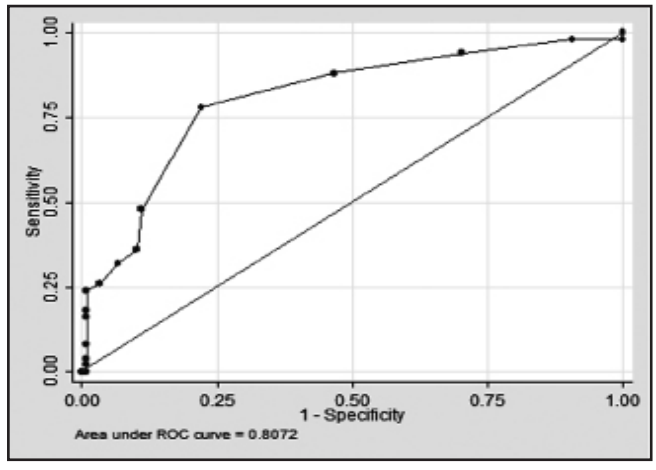

Fig 5: **Roc Curve for the Level at the Tunnel Inlet**

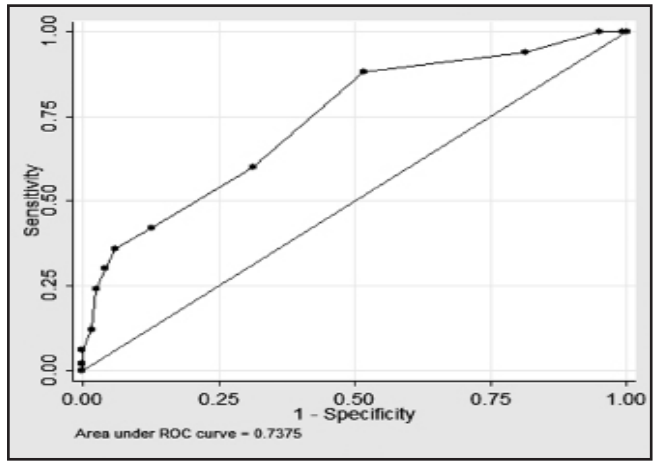

Fig 6: **Roc Curve for the Level of the Tunnel Outlet**

Based on the ROC curve, the cut-off for CSA for CTS in patients were 0.09 cm^2 at the level of the distal radio-ulnar joint (sensitivity 68% and specificity 68.64%), 0.10 cm^2 at the level of the inlet of the carpal tunnel (sensitivity 78% and specificity 77.97%), and 0.08cm^2 at the level of the outlet of the carpal tunnel (sensitivity 72% and specificity 55.93%).

The sensitivity of ultrasound to diagnose CTS by the increase in CSA at the level of the distal radio-ulnar joint for group 1 (mild to moderate CTS on NCS) is 58.54% and that for group 2 (severe CTS) is 66.67%. The specificity of ultrasound at this level compared to the electrodiagnostic study was 68.64%. The sensitivity of ultrasound to diagnose CTS at the level of inlet to the carpal tunnel (level of pisiform) in group 1 (mild to moderate CTS on NCS) was 78.05% and that for group 2 (severe CTS) was 77.78%. The specificity as compared to the electrodiagnostic study at this level

was 92%. The sensitivity at the level of the outlet of the carpal tunnel (level of hook of hamate) in group 1 (mild to moderate CTS on NCS) was 70.73% and that for group 2 (severe CTS) was 77.78%. The specificity compared to the electrodiagnostic study at this level was 66%.

The ability of the USG to pick up an increased CSA at any level according to the severity of the disease was 86.20% for group 1, and for group 2 was 85.7%

The mean of flattening ratio for the combined group at the level of proximal inlet was 3.06 with standard deviation of 0.75, at the inlet the mean was 2.74 with standard deviation of 0.58 and at the tunnel outlet it was 3.22 with standard deviation of 0.78. No significant correlation could be established between the flattening ratio and disease condition.

The positive predictive value and the negative predictive values of ultrasound vs. nerve conduction studies were calculated at each level.

1. Proximal inlet of the carpal tunnel
   Positive predictive value = 45
   Negative predictive value = 80
2. Inlet of the carpal tunnel
   Positive predictive value = 60
   Negative predictive value = 90
3. Outlet of the carpal tunnel
   Positive predictive value = 41
   Negative predictive value = 83

Discussion:

The clinical diagnosis of CTS usually relies on typical signs and symptoms which are followed by electrodiagnostic studies for confirmation. Stand-alone signs and symptoms have shown to limit diagnostic accuracy, while electrodiagnostic study cause discomfort, is time consuming, expensive and not widely available. Electrodiagnostic studies demonstrate the physiological malfunctioning of the median nerve while ultrasonography picks up the structural abnormalities.

In literature, US measurement used in CTS diagnosis is the cross-sectional area of the nerve at various levels of the carpal canal, the flattening ratio, the swelling ratio, and the increased palmar bowing of the flexor retinaculum. In some studies cross-sectional area was performed at a single level [4,5, 27-30] mostly at the proximal carpal tunnel. In several studies CSA was measured by ellipsoid formula [10, 28, 31] but a more accurate measure is obtained by using continuous boundary trace of the nerve, because the nerve does not always have a perfect ellipsoid shape, which the method used in our study. Nakamichi and Tachibana[11] directly compared the measurements of the median nerve obtained sonographically with the measurements found in anatomical cross-sections in cadaver limbs. Ultrasound is a precise method for determining these measurements, later confirmed by Kamolz *et al*[32].

Kamolz *et al*[33]. stressed the need for standardisation of the median nerve CSA cut-offs. In our study, the cut off for an abnormal nerve was taken as 0.09 cm^2 at the level proximal to the inlet, 0.10 cm^2 at the inlet and 0.08 cm^2 at the tunnel outlet with sensitivities of 68%, 78% and 72% respectively and specificities of 68.64%, 77.97% and 55.93%; also the cross-sectional area at inlet of the carpal tunnel showed higher sensitivity and specificity (78% and 77.97%) for CTS as compared to the proximal inlet and outlet of the tunnel. The majority of the studies published previously had values ranging from 9mm^2 to 12 mm^2 in different populations, which corroborates with our findings also.

In our study, the sensitivity and specificity of the flattening ratio was not calculated, as it did not show any significant correlation with the presence of the disease. However, the mean of the flattening ratios for the cases and the controls was taken separately.

Ultrasonography is useful in CTS diagnosis, providing anatomic images of the median nerve, neighbouring structures, and mass-occupying space in the carpal canal. The advantages of ultrasonography is that it is low cost, takes a shorter duration to perform the investigation compared to nerve conduction studies and it is more commonly available, besides it is painless and non-invasive; and gives dynamic images. US is operator dependent, but shows high reproducibility after adequate training of the operators[34].

The sensitivity of ultrasound to detect CTS by the increase in the cross-sectional area of the median nerve as compared to the nerve conduction studies is 90% with the US value being positive at any one anatomical level. The specificity for this is 45%. According to this study, based on the sensitivity, the best level to look for the compression of the nerve by increase in the cross-sectional area is at the level of the carpal tunnel inlet, however a combination of the CSA at the inlet and outlet of the carpal tunnel will improve the screening accuracy of the test.

Conclusions:

Ultrasonography is useful in CTS diagnosis, providing anatomic images of the median nerve, neighbouring structures, and mass-occupying space in the carpal canal. The advantages of ultrasonography is that it is low cost, takes a shorter duration to perform the investigation compared to nerve conduction studies and it is more commonly available, besides being painless, non-invasive and gives dynamic images. US is operator dependent, but shows high reproducibility after adequate training of the operators[34]. Therefore in conclusion as ultrasonography is more widely available as compared to NCS, is non- invasive with the added benefit of being cost-effective, it can be used as a good screening tool for the diagnosis of CTS. This investigation becomes especially useful when the availability of nerve conduction studies is difficult.

Future Directions:

For further studies, it may be useful to look for the cross-sectional area of the median nerve at the 3 different levels (at the distal radio-ulnar joint, the level of the pisiform and at the level of the hamate) 3-6 months post surgery or conservative management along with concomitant nerve conduction studies and compare with the pre-operative values. This may be especially useful in those patients who continue to have symptoms despite treatment especially surgical management.

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