DIASTEMATOMYELIA (SPLIT CORD SYNDROME): A RETROSPECTIVE STUDY

Samir H. Abood *FRCS,* **Abdul Amir Jasim** *FICMS,* **Sarmad A. Ibrahim** *FICMS*

Abstract

Background: Diastematomyelia is the term used to describe the malformation in which the spinal cord is split into two hemi cords, by a bony or cartilaginous ridge, each has a single set of dorsal and ventral nerve roots and is contained within its own dural sheath. On the other hand, diplomyelia indicates that the two spinal cord segments are completely duplicated; four sets of dorsal and ventral nerve roots are present within a single dural sleeve.

Objective: To study the clinical, radiological and pathological patterns of diastematomyelia in Iraq, and to emphasize the importance of thorough radiological investigations in order to achieve adequate surgical treatment**.**

Methods: Five patients of proved diastematomyelia were studied retrospectively regarding age, sex, and clinical and radiological features. Four of them were treated surgically by removal of the ridge and release of the tethered cord.

Results: The patients were two male and three females. Four of them were below one year of age. Back deformity was the main presenting feature in the first few months of life, while delayed walking was the main presentation after one year of age. The commonest pathological findings were split cord with bony ridge associated with lipoma. CT and MRI provided excellent anatomical and pathological details. Two of the patients developed CSF leak following surgery and had to be reopened. All children made good recovery**.**

Conclusion: Closed spinal midline developmental defect occurs more readily in girls than in boys. Multiple and complex abnormalities are observed: diastematomyelia, diplomyelia, and vertebral malformations. There are three groups of structures involved in the pathological process, skin, bone and neural tissues. CT and MRI provide excellent radiological diagnosis, and should obviate the need for myelography in this group of patients. Surgical treatment is the only way to correct the multiple deformities and defects, and to release cord tethering.

Key words: Diastematomyelia, Diplomyelia, Split cord malformation (SCM), Tethered cord syndrome, Bony cartilagenous spur, Fibro cartilagenous spur, Lipoma, Meningomyelocele.

Iraqi J Med Sci, 2004; Vol. 3 (1): 62-67

Introduction

Diastematomyelia (Type I SCM) is the term used to describe the malformation in which the spinal cord is split into two hemi cords and each has a single set of dorsal and ventral nerve roots and is contained within its own dural sheath, separated by a dural-sheathed rigid osteocartilagenous (or bony) median septum. The name is derived from a Greek word meaning separation and marrow (i.e., spinal cord). On the other hand, diplomyelia (Type II SCM) indicates that the two spinal cord segments are completely duplicated; four sets of dorsal and ventral nerve roots are present within a single dural sleeve and separated by a non rigid fibrous median septum[1]. More recent investigations have revealed that diastematomyelia and diplomyelia represent opposite ends of a continuum with few, if any, "true" examples at either end[1]. Because this group of malformations appears to have a common embryological disorder, Pang *et al*[1] made a good argument that the terms diastematomyelia and diplomyelia should be abandoned in favor of calling these lesions collectively split-cord malformations (SCM).

The error in embryological development that produces split-cord malformations is subject to continuing debate. Where as failure of primary neurulation can explain dorsal abnormalities regardless of whether they are open or closed, a ventral disruption in normal spinal cord formation is needed to produce a split-cord malformation. One theory that unifies the spectrum of this group of neural tube defects (NTD) states that the disordered mechanism occurs during gastrulation, when all three germ layers are in proximity[1,2]. Investigators presume that a second neuroenteric canal becomes invested with mesenchyme to form an endomesenchymal tract that splits the notochord and neural plate and that the timing and

Dept. Surgery-Divsion of Neurosurgery, College of Medicine, Al-Nahrain University.
Received 8th March 2003: Accepted 19th January 2004.
Address Correspondence to Dr. Samir H. Abood.

severity determine the extent of the resultant malformation. Although often the two hemicords are approximately the same size, significant discrepancies can occur. The spinal cord often reunites distal to the cleft. Also, two separate areas of clefting have been reported. Sometimes a syrinx can be seen and is usually confined to one of the hemicords (When two dural sheaths are present, a bony or fibrocartilaginous spur exists at the caudal end of the cleft and may be attached to the surrounding bone dorsally, ventrally, or both. The spinal cord is tethered by this spicule as well as medially to the dura mater surrounding the spur. Frequently, the conus medullaris is low lying and tethered by a thickened filum terminale. Other NTDs, especially myelomeningoceles and neurenteric cysts, can be seen along with split-cord malformations and, a bony spur also would be expected in the presence of a hemimyelomeningocele, which combines an open NTD and a closed NTD. The anterior component of this embryological disorder results in a high incidence of bony vertebral column abnormalities that include failure of vertebral body segmentation; hemivertebrae; butterfly vertebrae; a widened spinal canal diameter, fusion of the pedicles, transverse processes, and laminae.

The clinical presentation of diastematomyelia is related to the group of tethered cord malformation as listed in table (1)[3,4].

Table 1: Clinical presentation

Parameter	Children	Adults
Pain	Uncommon complaint: when present, is usually in back and legs, anal-perianal pain is not noted.	Present & most often localized to anus & perineum: diffuse & bilateral, occasional shock like sensation
Foot deformity	Common: early sign of neuromuscular imbalance: usually progressive cavovarus deformity.	Not seen as a presenting or progressive symptom.
	Common: usually worsening scoliosis.	
Progressive spinal deformity	Present in most as walking difficulties, gait abnormalities, and regression in gait training.	Not seen as a presenting or progressive symptom.
Urological symptoms	Common: usually as absence of dry periods between diapers. Continuous drippling, delayed toilet training, recurrent infection and enuresis.	Also common: urinary frequency, urgency, felling of incomplete voiding, stress incontinence, overflow.
Trophic ulceration of the legs and feet	Relatively common	Uncommon
Cutaneous stigmata of dysraphism	Common	< 50% of adults have external signs of spinal dysraphism
Aggravating factors	Growth spruts	Many factors precipitate acute symptom onset: trauma, maneuvers associated with starching of conuc, spinal stenosis.

Treatment is usually surgical aiming at eliminating the septum and release the tethered spinal cord. The absolute surgical indication for diastematomyelia is the existence of a progressive neurological deficit, and as a preliminary to the surgical correction of scoliosis. Other indications are relative, treated expectantly; many children with diastematomyelia who are initially asymptomatic eventually require operation on account of backache, painful or spastic lower limbs, or sphincter troubles[4]. Neurological improvement following an operation is usually modest and is restricted to symptoms and signs of recent onset. Children with dysraphism never improve beyond their degree of congenital neurological deficit. Recurrence of symptoms due to secondary adhesions of the spinal cord to the dura at the site of the operation is an uncommon late complication[5].

Aims: To study this congenital deformity regarding: the clinical presentation, pathological findings, radiological features, and the technique and outcome of surgical treatment.

For many years complicated cases of NTD were treated surgically without adequate radiological investigations, due to the unavailability of CT and MRI. Adequate myelographic study is difficult to obtain in this group of patients because of their age and the often infected back deformity. Consequently no details of anatomy were available during surgery, which often consisted only of obtaining skin cover of the defect.

Material & Methods

The material of this study included 5 patients with diastematomyelia .The patients files were reviewed and all biographic, clinical, radiological, and treatment data uniformly collected according to previously designed data sheet. The collected data includes; patients age, gender, duration of illness, the presenting symptom, and physical signs.

The diagnosis depended on CT with bone study in the axial and sagital planes, and MRI. Patients suspected to have SCM and recommended for radiographic examinations included patients with: signs and symptoms suggestive of tethered cord syndrome, cutaneous stigmata of occult spinal dysraphism without neurological symptoms and

history of an open myelomeningocele repaired at birth with recent progression of neurological deficits. The radiological findings by spiral CT scan and MRI are seen in figure 2, such as diastematomyelia associated bony or fibrocartilaginous spur, lipoma and myelomeningocele were also recoded.

Surgical technique[4]:

The aim of surgical treatment is to eliminate the septum, and release the spinal cord. In type I (SCM), the bony septum is always enclosed with in a dural sleeve. The bone is frequently fused with the neural arch dorsally. A laminectomy is performed carefully around the attachment of the septum until only a small island of lamina is left attached to the dorsal end of the septum. This permits subperiosteal dissection of the septum from its dural sleeve deep within the median cleft. Once the dorsal attachment of the septum is eliminated, it is no longer rigidly anchored at both ends, and can be resected.

All other tethering and associated lesions such as thickened filum terminale, lipoma, dermal sinus tract, and adhesions from previous surgery were also treated.

The operative data were documented including the surgical finding, the extent of spur resection and freeing of the tethered cord. Follow-up notes were reviewed and the patient's clinical findings were documented. The outcome of treatment was considered good if the patient has stable neurological function and poor if he is worse for any reason.

Results

Tables 2, 3, 4 & 5 summarize the age and gender distribution, the clinical, radiological, operative findings and complications: respectively. The back deformity is usually in the form of cystic swelling, partially covered by skin. Sometimes skin pigmentation or a tuft of hair is seen (Figure 1). Foot deformity a talipes equino varus, usually involves one leg (Figure 2).

Table 2: Age & sex distribution and presenting symptoms

Age	Sex	Presenting symptom	Duration
3 months	Female	Low back deformity (Myelomeningocele)	Since birth
7 months	Female	Low back deformity (Meningocele)	Since birth
7 months	Male	Low back deformity (Meningocele)	Since birth
24 months	Male	Limping	1 year
60 months	Female	Limping	6 months

Table 3: Clinical Features

Symptom	< 1 year	1-2 year	> year
Pain	No	No	No
Foot deformity	Yes (Rt. Foot)	Yes (Rt. Foot)	Yes (Rt. Foot)
Progressive spinal deformity	Yes (Kyphoscoliosis)	No	No
Motor deficit	Yes (Rt. Foot)	Yes (Rt. Foot)	Yes (Rt. Foot)
Urolog. symptoms	?	No	No
Trophic ulcers	No	Yes (Rt. Foot)	No
Cutaneous stigmata	Lumber or lower dorsal meningocele with assoc. tuft of hair	Lower lumber tuft of hair & skin pigmentation	Lower lumber skin dimple associated with tuft of hair
Aggravating factors	No	Growth spurt	Growth & activities

Table 4: Operative findings

Pathology	No.
Bony ridge	3/4
Fibrocartligenous ridge	1/4
Associated lipoma	3/4*

* one still not operated on

Table 5: Operative Complications

Complication	No.
Cerebrospinal fluid leak	2/4*
Hydrocepalous	0
Wound infection	0

* both were re-explored, one required lumbo-peritoneal shunt

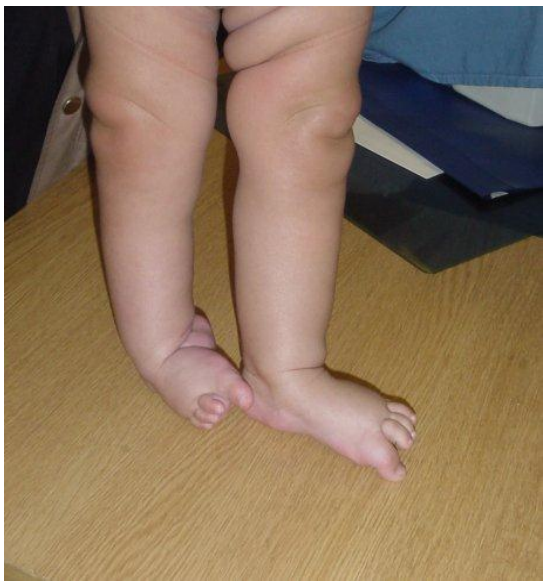

Figure 1: Foot deformity in a 7 months old child

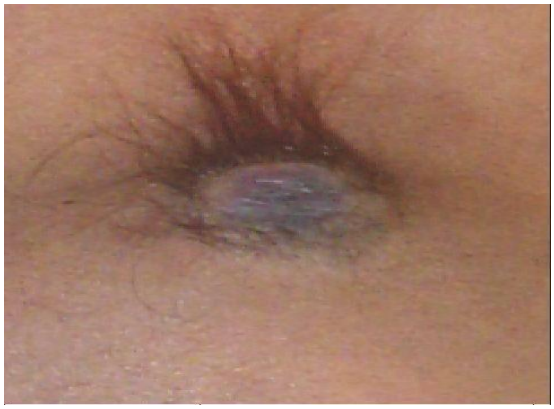

Figure 2: Back deformity, cystic swelling and tuft of hair in the lumbosacral region of a 7 months old child

Radiological findings:

Figures 3-6 of spiral CT and MRI clearly demonstrated the split cord, the bony spur, associated lipoma and other evidence of cord tethering.

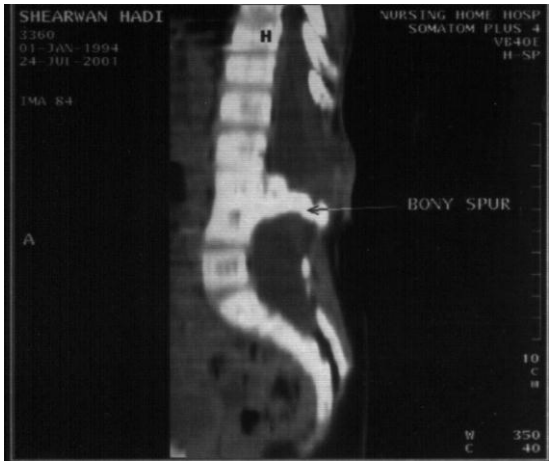

Figure 3: Spiral CT. Sagital reconstruction showing the bony spur attached to the back of the vertebrae

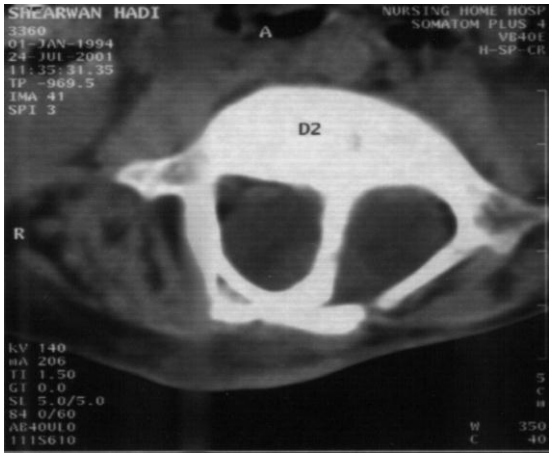

Figure 4: Spiral Ct. Axial view: showing the complete division of the spinal canal by the bony spur

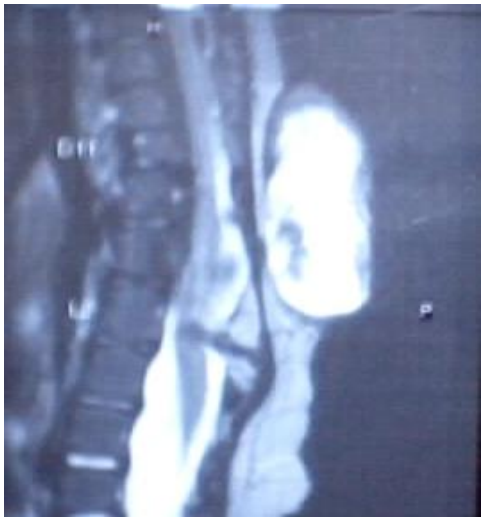

Figure 5: Sagital MRI showing the bony spur, and an associated lipoma

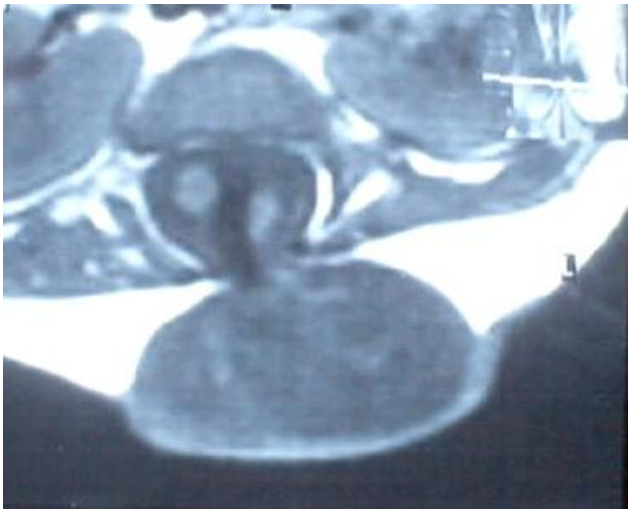

Figure 6: Axial MRI showing diastematomyelia, the Rt. Is closed and the Lt. is open in to the cyst

Follow up:

All children who were operated on made good postoperative recovery. Three of the patients were followed up for six months, two had CSF leak and had to be re-operated. The result of surgery was considered good in the three cases. The two eldest children showed improvement in gait and foot deformity.

Discussion

Diastematomyelia is an uncommon dysraphic state usually seen in infants and young children, and is a rarity in adults[4,6]; only 19 reports have been published in the literature so far. In this study all patients were below 5 years of age. The clinical manifestations of split cord malformation are

similar to those seen for other forms of closed neural tube defects. Split cord malformations are seen approximately twice as often in females[6,7,8] which is some what similar to the results in this study, where females comprised 60% of patients. In children the onset of symptoms is usually insidious, only rarely a definite precipitating event is recognized. Depending on the age of the child and the location of the split cord lesion, the signs to look for are definite leg or arm weakness, deteriorating gait or regression in gait training in toddlers, and decreased spontaneous movement in the lower extremities in infants[6,7]. In this study three patients below one year (60%) were accidentally found to have split cord lesion in association with myelomeningocele, another two patients (2, and 5 years old) presenting as delayed walking and limping respectively. Trophic changes of the lower extremities from sympathetic denervation, for example smooth shiny skin; hair loss, nail changes, and non healing ulcers in the toes are seen occasionally in older children[4,6,7]. In this study one patient, two years old had trophic skin ulcer in the leg.

Foot deformity most likely results from neuromuscular imbalance at a time when the tarsal, metatarsal, and phalangeal bones are actively growing in early childhood, and aligning with each other along closely set joint surfaces[9]. In this study more than 1/2 of the cases had foot deformity.

Almost all children with tethered cord syndrome have some cutaneous stigmata of underlying dysraphism, but less than 50% of adult do. Midline hairy patches are highly correlated with split cord malformation[4,6-8]. Two clinical points need to be emphasized even though type II split cord malformations do not present a dramatic radiographic picture as type I malformation[10-13], a stiff fibrous septum was found in all type II lesion explored. Usually in the symptomatic group of patients, there were almost equal number of patients having type I and II lesion[8]. In this study bony septum (type I) was found in three patients and fibro cartilaginous septum in two with associated lipoma in three patients.

Postoperative CSF leak was seen in two cases, this was due to dural tear during dissection of the dura from the bony spur. In this study two cases of CSF leak were re explored, and one required lumboperitoneal shunt. In both patients the leak stopped. Thus, this study strongly argues that both types of SCM are cord tethering lesions likely to cause neurological damage, and both should be treated. All Type II SCMs should be explored, even if imaging studies did not reveal a definite median septum. The entire neuraxis should be studied to look for other tethering lesions, which should also be treated. Surgery is excellent for improving or stabilizing the neurological status.

The association between SCM and opened myelomeningocele has been shown to be between 26 and 80 percent[12,14-16] and the wider use of screening MRI on children with myelomeningocele will be likely to turn up with even more cases of SCMs before they become symptomatic.

Conclusion

1. The signs and symptoms of SCM are similar to those described for the tethered cord syndrome. Children commonly present with gait disorder and less so with pain and progressive spinal and foot deformities.
2. MRI and CT scan are useful screening tests for SCMs in delineating anatomical details of the splitcord lesion and in predicting associated lesions. With these two tests, all SCMs can be accurately classified into Type I or II lesions preoperatively, with no crossover ambiguities.
3. Even with the most sophisticated CTscan and MRI, many Type II SCMs will not be shown radiographically to possess a definite fibrous septum even though one is always present to tether the hemicords. As with Type I lesions, all Type II SCMs should be surgically explored regardless of whether a definite septum is found on neural imaging.
4. Although longer follow up is needed. The surgical outcome for SCM appears to be excellent.

Recommendation

All patients with suspected spinal dysraphism, should have full radiological evaluation with spiral CT and MRI in order to discover SCM early. For better evaluation of the result of surgery, electro-physiological and urological studies and longer follow up is required.

References

1. Pang, D., Dias, M.S., and Ahab-Barmada, M.: Split cord malformation. Part I: A unified theory of embryogenesis for double spinal cord malformations. Neurosurgery, 1992; 31: 451-80.
2. Dias, M.S., and Walker, M.L.: The embryogenesis of complex dysraphic malformations: a disorder of gastrulation? Pediatric Neurosurg, 1992; 18: 229-53.
3. Pang, D., and Wilberger, J.E.Jr.: Tethered cord syndrome in adult. J Neurosurg, 1982; 57: 32-47.
4. Pang, D., Tethered cord syndrome: Neurosurgery/editors, Wilkins, R.H., and Rengachary, S.S., Mc Grow-Hill. New York 2nd edition; Vol.III: Part XI, 1996; pp 3465-97.
5. Guthkelch, A.N.: Diastematomyelia: Neurosurgery/editors, Wilkins, R.H., and Rengachary, S.S., Mc Grow-Hill. New York 2nd edition; Vol.III: Part XI, 1996; pp 3505-9.
6. Wolf, A., Bradford, D., Lonstein, J., Ogilivie, J., and Erickson, D.: The adult diplomyelia syndrome: Spine, 1987; 12(3): 233-7.
7. Benzerki, L., Jarmmouni, R., Zouhair, K., and El-Quazzani, T.: Closed spinal dysraphism. Apropos of 3 cases: Ann Dermatol Venereol, 1996; 123(6-7): 378-81.
8. Pang, D.: Split cord malformation. Part II: Clinical syndrome. Neurosurgery, 1992; 31: 481-500.
9. Sattar, M.T., Bannister, C.M., and Turnbull, I.W.: Occult spinal dysraphism-the common combination of lesions and the clinical manifestations in 50 patients. Eur J Pediatr Surg, 1996; 6 (Suppl 1): 10-14.
10. Herren, R.Y., and Edwards, J.E.: Diplomyelia. Arch Pathol, 1940; 30: 1203-14.
11. McCaig, C.D.: Myoblasts and myoblast-conditioned medium attract the earliest spinal neurites from frog embryos. J Physiol (Lond), 1986: 375: 39-54.
12. Meacham, W.F.: Surgical treatment of diastematomyelia. J Neurosurg, 1967; 27: 78-85.
13. Scotti, G., Musgrave, M.A., and Harwood-Nash, D.C.: Diastematomyelia in children: metrizamide and CT metrizamide myelography. Am J Roentgenol, 1980; 135: 1225-32.
14. Moes, C.A.F., and Hendrick, E.B.: Diastematomyelia. J Pediatr, 1963; 63: 238-48.
15. Till, K.: Spinal dysraphism: a study of congenital malformations of the back. Dev Med Child Neurol, 1968; 10: 470-7.
16. Prasad, V.S., Sengar, R.L., Sahu, B.P., and Immaneni, D.: Diastematomyelia in adults. Modern imaging and operative treatment. Clin Imaging, 1995; 19(4): 270-4.