Spinal dysraphism controversies: AIIMS experiences and contribution

A. K. Mahapatra
Department of Neurosurgery, All India Institute of Medical Sciences, New Delhi, India

ABSTRACT

A large series of split cord malformation (SCM). Over the last 22 years, we have operated more than 1500 patients of SD, of which over 450 are (SCM), and 300 are with various lipomatus malformations. About 55% type II and 45% type I SCM. A separate sub-classification of type I SCM (a,b,c and d), is presented which alter the surgical approach and influence the results. Overall improvement following surgery in patients with SCM was observed in 94%. Fifty percent patients improved and 44% remained stable. However, deterioration was noticed in 6%, mostly with composite type of SCM. A paradigm for management of SCM is provided taking into consideration also the author's large experience.

Key words: Split cord malformations, spinal dysraphism, surgery

INTRODUCTION

Spinal dysraphism is an uncommon condition reported in 30 to 40/100000 line birth.[1,2] The incidence has significantly decreased over the last 5 decades, all over the world; however, fall in incidence in much lesser in developing countries with poor socio-economic status, educational background and financial condition of general population.[3-5] In India spinal dysraphism (SD) is still a major public health problem with added problems resulted from lack of medical personnel, be it pediatrician, neurologist or neurosurgeon and also the lack of diagnostic facilities. In this presentation, I have highlighted the management problems, controversies and more importantly logistics of management of these patients.[3,5]

MATERIALS AND METHODS

AIIMS is a referral center and center of excellence. Over the last 22 years, we have operated more than 1500 patients of SD, of which over 450 are split cord malformations (SCM), and 300 are with various lipomatus malformations [Table 1]. Others included, tethered cord syndrome (TCS), meningomyelocele (MMC) and myelocystoceles. A significant number of patients operated in neonatal period, need reoperation as many pediatric surgeons and general surgeons do operate these patients without MRI; hence, they usually miss the underlying various associated pathologies.

Management Protocol at AIIMS

All the children were evaluated clinically in out patients department (OPD). MRI of brain and spine carried out and CVJ evaluated to establish the associated chiarimalformations. Renal function was evaluated to find out the association of urogenital anomalies.Chest X-ray was done to find out lung, rib and diaphragmatic abnormalities and associated dexocardia. Patients were subjected to surgery and post operatively nursed in prone position. Acetazolamide was routinely used. Most of the patients were discharged between 7th and 10th post-operative days. Post-operative wound break down, CSF collection and CSF leak were managed according to their own merit and patients were discharged when they were fit. Patients were followed up in OPD. In patients with hydrocephalus VP, shunt was inserted

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Table 1: Profile of spinal dysraphism at AIIMS

No. of patients	%
SCM	400
Lipoma	250
Dermal sinus	65
Others	785
Total	1500

(1989-2010)

Address for correspondence: Prof. A. K. Mahapatra,
Department of Neurosurgery, All India Institute of Medical Sciences, New Delhi, India. E-mail: akmahapatra_22000@yahoo.com

in the same sitting with surgery for spinal dysraphism [Table 2].

Management of SCM 400 Patients

Over the last 22 years, we have managed over 400 SCM, which include 55% type II and 45% type I SCM. Over the last 8 years we are also following our sub-classification of type I SCM (a,b,c and d), which alter the surgical approach and influence the results.There were 7 patients with posterior spur, where the spurs were arising from the lamina rather than from the posterior surface of the vertebral bodies. In posterior spur, laminotomy is difficult to perform; hence, spur were separated from lamina before the laminotomy was performed, to avoid the dural tear and damage to the spinal cord.Intravenous methyl-prednisolone was used during surgery when excessive handing of spinal cord was undertaken at the time of surgery [Table 3].

Results of Surgery in SCM

Overall improvement following surgery in patients with SCM was observed in 94%. Fifty percent patients improved and 44% remained stable. However, deterioration was noticed in 6%, mostly with composite type of SCM. None of the patients operated prophylactically had neurological deterioration [Table 4].

Management of Patients with Lipoma

Over the last 22 years, 250 patients were operated upon. For this presentation, only 140 patients were analyzed in detail [Tables 5 and 6]. Type of lipomasareenumerated in Table 5. The commonest lipoma was conuslipoma followed by filumlipoma and lipomyelomeningocele was recorded in 32 patients.The MRI scan in patients with lipoma revealed large number of associated pathologies [Table 6] more common among them were SCM and syringomyelia recorded in 12% each and developmental tumor like dermoid and epidermoid in 9%. Hydrocephalus was relatively rare in lipomas, and was noticed in only 6.3%. Among 60 patients in whom detailed urogenital evaluations were carried out, 25% had abnormalities in the form of horse shoe kidney, single kidney, atrophic and ectopic kidney. Hydronephrosis was noticed in 6 patients (10%). Our findings justify the detailed evaluation of kidney function.

Surgery and Outcomes

All the patients were subjected to surgery, and this included patients who had no neurological deficits. Patients were operated with long vertical skin incision and liberal laminotomy to expose the entire extent of thelipoma and also the filum. Maximal excisions of lipomas were carried out keeping in mind the higher risk of deterioration in complex and transitional conuslipoma.

Table 2: Management protocol at AIIMS

Clinical evaluation
Assessment of kidney function
MRI of spine and brain
MRI of spine and brain CT
Surgical intervention
Patients were offered prophylactic surgery even though no neurological deficit
Post-operative nursing in prone position for 10 to 15 days
Regular follow-up at OPD

Table 3: SCM types and cause of tethering

SCM	Median roots
SCM	Thick filum
Composite type	Bony or fibrous septum
Multiple site of split	Extradural or intradural bands
Presence of posterior spur	Dermal sinus
Neuroenteri cyst	
Lipoma	

SCM: Split cord malformations

Table 4: Results of surgery overall results of 400 patients with spinal dysraphism

	%
Result of surgery in patients with SCM (400 patients)	
Overall improvement	50
Patients remained stable	44
Patients deteriorated	06
Results in spinal lipoma (250 cases)	
Overall improvement	90
Motor improvement	34.6
Sensory improvement	48.6
Improvement in urinary problem	25.0

Table 5: Location of lipoma in 141 patients

Locations	%
Intradural	22
Intradural, intramedullary	46
Conus lipoma (dorsal type)	34
Conus lipoma – transitional type	08
Filum lipoma	31
Lipo-myelomeningocele	32

Table 6: MRI findings in patients with lipoma associated pathology

	%
SCM	12
Epidermoid/dermoid	11
Neurenteric cyst	4
Syringomyelia	12
Hydrocephalus	6.3
Chiari malformation	7

SCM: Split cord malformations; (141 patients)

Filumlipomas were totally excised. In patients with conuslipoma, after maximal excision of the lipoma, conus was reconstituted by applying 4 to 5, 5 to '0' *Vycril* suture and dura was meticulously closed. In situation when dura was deficient, lumbar fascia was used as the dural graft. Methylprednislone was prescribed to patients in whom conus was handled for long duration and deterioration was suspected. Post-operatively patients were nursed in prone position, i.e., in a slight head down position. The patients also received antibiotics, acetazolamide and analgesic syrup for 5 to 10 days, depending on the case to case basis.Most of the patients were discharged between 7th and 10th postoperative day. Patients were regularly followed up in neurosurgery OPD.

Overall improvement or stabilization was observed in 90%. In 10%, deterioration was recorded [Table 4b].Motor and sensory improvement was recorded in 34.6 and 48.6% respectively. Some improvement in urinary symptoms was recorded in 25% of patients. Among the 10% of patients who had deteriorated, approximately 50% of them had some improvement in long term follow up [Table 7].

Table 7: Overall complications (141 patients)

	%
Wound infection	5
CSF leak/collection	15
UTI	10
Deterioration of deficit	6
Death	2

Table 8: Factors influencing the outcome

Age of the patients
Neurological status
Nature of the pathology
Associated pathologies and hydrocephalus
Cranial anomalies
Other system anomalies

DISCUSSION

Incidence of SD in the US, UK and other developed countries has significantly decreased. However, it still remains an important public health problem in developing countries like India.Approximately 8 to 9/10000 live births does have some form of SD in India. Every year, we come across over 300 casesin our OPD, which is a very large number.However, because of lack of beds; only 100 to 120 patients get admission for surgical repair.

Prenatal diagnosis is an important aspect to diagnose these lesions.Unfortunately, due to lack of awareness and infrastructure, large number of patients during pregnancy do not get antenatal care, and hence, no USG evaluation of the fetus is done. Medical Termination of pregnancy (MTP) in India has been legalized since 1974. If diagnosed antenataly, MTP should be considered in early stage of pregnancy.

Factor Influencing Outcome

Large number of factors influence the outcome,[6-17] few among them are age of the patient, type of pathology, cranialanomaly, presence of hydrocephalus, paraplegia, degree of neurological deficit and associated vertebral and systemic anomalies.Some patients do have situsinversus ordextrocardia.[6,7]In our study 3 patients had situsinversus, and one patient had eventration of diaphragm [Table 8].

Controversies: Related to Management

As large number of factors influence the outcome,[6,16,17] the result can be dependent on many factors, hence, the controversies. Usefulness of performing investigations like urodynamic studies is controversial[3,5,13] which is by and large helpful in assessing the bladder function, however, it's role is limited in children below two years of age.

Another controversy is regarding surgery in paraplegic children. Majority of pediatric and neurosurgeons do not recommend surgery in paraplegic children. However, as a policy we in our department do not refuse the surgery to a paraplegic baby with MMC or any other form of SD, in case the family agree for surgery, fully understanding the long term implications and outcome of paraplegic baby, who otherwise have very little chance of improvement of power in lower limb power.[3,4,5,18,19] We had reported, two infants recovering from paraplegia following surgery for neuroenteric cyst.[20,21]

Significant controversies revolved around the surgery in asymptomatic patient, be it lipoma, meningocele or SCM.[2,12,22,23] Pierre Kahn *et al.*, in 1997, had suggested surgery for all lipoma, but subsequently Kulkarni *et al.* in[24] 2004, from the same center, recommended conservative management of spinal lipoma. Pang *et al.*[12] repeatedly have suggested prophylactic surgery for spinal lipoma. However, in their opinion surgeon should have sufficient experience to deal with complex problem like spinal lipoma. In an another publication Dorward *et al.*[2] had published their experience of prophylactic surgery in lumbosacral lipoma, and after detailed analysis of pitfalls they concluded in favor of prophylactic surgery.In our Institute we have observed 10% patients coming in 3rd and 4th decade with neurological deterioration and even with chronic renal failure with recurrent urinary tract infection. As a policy we in our Institute recommend prophylactic surgery [Table 9].[3,4,17,18,25-28]

Table 9: Controversial issues in management

Operate or not a paraplegic child
Urodynamic study in babies less than two years
Surgery for asymptomatic patients
Extent of excision of conus lipoma
Surgery for multicentric dysraphism
Timing of VP shunt/scoliosis surgery

Extent of excision of lipoma is controversial. It can vary from partial excision, to subtotal and total excision depending on type, size and complexity of conuslipoma[2,5,10,11,12,22,29-31] There is very little debate regarding total excision of filumlipoma, which is usually easy to remove.[5,13] In case of conuslipoma near total or total excision is possible.[5,12,18,22] We also believe in near total or total excision of conuslipoma.[5,18] However, in transitional type of conuslipoma or complex type of conuscaudalipoma, when large number of nerve roots are buried in lipomatous tissue or when lipoma encircles the conus and distorts the anatomy, a relatively conservative surgical excision is justified to avoid the post operative deterioration.[12,18,30]

A small percentage of patients may have tethering at multiple sites.[10,11,29,32,33] Rarely, patients may have 3 or 4 level spinal dysraphism comprising SCM, lipoma, neuroenteri cyst, dermal sinus and myelocystocels.[10,27] This type of lesion also results in controversies in various hypothesis in neural tube closure, which are zip theory versus multisite closure theory.[28,32,33] In our publications we favored the multistage closure theory.[33] Multisite spinal dysraphism also triggers the controversies regarding the one stage or multistage surgery, due to the prolonged surgery when surgeon deals with 3 to 4 pathologies at various levels starting from cervical to lumbosacral area.However, in older children more than 2 to 3 years of age we have tried to deal all the lesions in a single stage.[10,27,28] Single stage surgery, cuts down cost and need for multiple admissions.

Hydrocephalus is present in 80 to 90% case of MMC as compared to 30 to 35% in lipoma and SCM. Presence of hydrocephalus is an important and independent prognostic factor for cognitive function. Hydrocephalus also may result in higher incidence of CSF leak or pseudomeningocele. Therefore, insertion of ventriculo-peritoneal shunt is more than desirable in these patients. There is very little literature available dealing with hydrocephalus as a separate entity. In our Institute, initially we used to insert VP Shunt as first stage and then repaired MMC or SD as second stage.This approach increases the hospital stay and also the hospital cost. Hence, over the last 10 to 12 years we are operating the patients in single stage.First shunt is inserted and in the same sitting the SD is operated so that the patient's family and the hospital get the benefit [Table 7].

No surgery is without a complication. Overall complication of surgery in SD can range from 15 to 20% and approximately 10% can have neurological deterioration.[2,3,5,12,18,22,25,30] This questions the need for prophylactic surgery.[2,24] In our experience neurological deterioration occurred in 6 to10% cases.[3,5,18,25] In our study of SCM, none of the patients undergoing prophylactic surgery had neurological deterioration,[3,4,25] however, 10% of patients with conuslipoma who underwent prophylactic surgery had postoperative deterioration, and 50% of them on follow up showed improvement.[5,18] Hence, we recommend conuslipoma surgery which must be undertaken by a neurosurgeon with experience in this type of surgery.

Spinal teratomas are rare condition, associated with spinal dysraphism.[8,34,35] We have operated more than 10 teratomas in last 10 years.[8,35] In one of our patient, a one-year-old child, the teratoma recurred in 1 year time and the patient needed repeat surgery. By and large recurrence is more frequent in immature teratoma as compared to matured ones.

Recently re-growth of excised bony spur is reported in the literature. However, such cases are very few and the natural history of these patients is not clearly known. In none of our cases, we had to re-operate a patient for re-growth of the spur.[36] Hence, long term careful follow up of SCM type I is necessary and incase of recurrence of symptoms, re-growth of spur must be considered as one of the possibilities.

CONCLUSIONS

Management of patients with spinal dysraphism is complex and large number of factors influences the outcome. Not surprizingly, a number of controversies revolve around the management of spinal dysraphism. We recommend brain and spinal MRI, and detailed urological evaluation of patients prior to surgery.We recommend prophylactic surgery and one stage surgery for multisitedysraphism. We also recommend one stage where VP shunt and surgery for spinal dysraphism are done together. Over the last two decades, we have collected world's largest series of SCM(400), Myelocystocele (32) and one of the large series of spinal lipoma (250 cases). We believe protocol-based management can lead to a very good result and minimize the postoperative neurological deterioration.

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