

Spina Bifida: Rehabilitation Problems in Rural Setting

Dr Ganesh Arun Joshi, MBBS, MD(PMR), DNB(PMR), Assistant Professor in PMR and Medical Education
Composite Regional Centre for Persons with Disabilities (CRC), Punarwas Bhavan,

Gopal Nagar, Khajuri Kalan Road, P.O. Piplani, Bhopal

Himanshu Dash, Dip. P&OE, Consulting P&O Engineer,

District Disability Rehabilitation Centre (DDRC) – Jashpur

F 18/1-2, Near Civil Hospital, Jashpurnagar (CG) 496331

Institution of study: District Disability Rehabilitation Centre (DDRC) – Jashpur

F – 18/1-2, Near Civil Hospital, Jashpurnagar (CG) 496331

Abstract

A follow up of four cases from a composite disability identification camp is presented here. The purpose of this case study is to reveal socioeconomic and access barriers for the children affected by spina bifida in the rural India. At the end are suggested some of possible ways and means to deliver rehabilitation services to them.

Key words – Spina Bifida, Myelomeningocele, Rehabilitation, Rural India

Introduction

Neural tube defects account for most of the congenital anomalies of the central nervous system. The development of neural tube is complete during first 3-4 weeks of pregnancy when pregnancy is not usually detected. Hence any assault during this phase may go undetected or forgotten. Neural tube defects present as spina bifida occulta, meningocele, myelomeningocele, encephalocele, anencephaly, dermal sinus, tethered cord, syringomyelia, diastematomyelia and lipoma involving the conus¹.

Spina bifida is of unknown origin with strong evidence to suggest mutations in folate responsive pathways. The management of this condition calls for a multidisciplinary effort² as well as effort from the society. The skin and bladder bowel problems remain un-noticed for long duration in these cases and hence remain unmanaged.

Case Report

District Disability Rehabilitation Centre, Jashpur conducted a composite disability identification camp for preschool and school going children (under 14 years) at

Bishrampur, Sarguja on 6th January 2006. Four (5%) out of 85 locomotor disabled children who attended the camp were suffering from myelomeningocele. All of them were given a visit at their residence by the authors to assess the social and architectural problems in living.

The cases attended the camp to receive solution for mobility (Table 1). Parents expected medical and surgical treatment in addition to assistive devices to make the child walk. Two older cases requested for treatment of pressure ulcers while the parents of the youngest were unaware of this complication. All except the youngest case requested treatment for urinary and fecal incontinence.

On examination, the younger two cases (Table 2) had lower neurological level at S3 while the older two cases had the neurological level higher up in lumbar spinal segments. All cases were incontinent in bowel and bladder. All of them had pressure ulcers. The elder two cases had most severe ulcers while younger two cases had less severe. Case no. 2 presented with the meningeal sac at 4 years age (Fig 1).

The medical rehabilitation services available to the cases (Table 3) were assessed. First two cases have access to a hospital at 1 km from their residence while the other two have access to a primary health centre at longer distances. All the cases except case no. 2 were operated for sac excision in neonatal phase. The surgical facility

Address for correspondence: Ganesh Arun Joshi, MD (PMR), DNB (PMR), Assistant Professor in PMR and Medical Education, Composite Regional Centre for Persons with Disabilities (CRC) – Bhopal, Punarwas Bhavan, Gopal Nagar, Khajuri Kalan Road, P.O. Piplani, BHOPAL (MP) 462021, Email – ganesharunjoshi@yahoo.com

Table 1 – History

<i>Case No.</i>	<i>Age</i>	<i>Sex</i>	<i>Presenting complaints</i>	<i>Expressed needs</i>
1	3	F	Inability to walk, dribbling of urine	Mobility
2*	4	F	Inability to walk, dribbling of urine	Surgery for lump in back, mobility, control of urine
3	7	F	Inability to walk, dribbling of urine, Non-healing ulcers	Treatment of pressure ulcers, mobility, control of urine
4	11	M	Inability to walk, dribbling of urine, Non-healing ulcers	Treatment of pressure ulcers, mobility, control of urine

* Elder brother had spina bifida

Table 2 – Examination findings

<i>Case No.</i>	<i>Sensory level</i>	<i>Motor Level</i>	<i>Bladder Bowel</i>	<i>Pressure ulcers</i>			
				<i>No.</i>	<i>Grade</i>	<i>Duration (years)</i>	<i>Management</i>
1	S3	L4 on left, above L2 right	Incontinent	1	1	1	Nil
2	S3	L4 on right, above L2 left	Incontinent	2	3	2.5	Dressing at home
3	T10	Above L2	Incontinent	7	4	4	Dressing at home
4	T10	Above L2	Incontinent	3	4	5	Dressing at home

Table 3 – Medical rehabilitation services received

<i>Case No.</i>	<i>Medical facility</i>	<i>Surgeries done</i>	<i>Assistive devices</i>	<i>Therapeutic exercises</i>	<i>Bladder-bowel management</i>	<i>Medical Expenditure till date</i>
1	Hospital at 1 km	MMC sac excised on day of birth at Bhilai. VP shunt, hernia and repeat surgery for meningocele done.	B/L KAFO Not in use	Few but regular	Tells about desire to void. Taken to toilet for bladder bowel training	Rs. 5,00,000
2	Hospital at 1 km	Not yet. Surgeon advised against surgery.	Nil	Nil	Nil	Rs. 2,000
3	PHC at 12 km	MMC sac excised on 8 th day after birth at Gwalior	Nil	Nil	Nil	Rs. 10,000
4	PHC at 2 km	MMC sac excised on day of birth at Bilaspur	Nil	Nil	Nil	Rs. 10,000

Table 4 - Social support

<i>Case No.</i>	<i>Occupation of head of family</i>	<i>Education of head of family</i>	<i>Annual family income</i>	<i>Family efforts</i>	<i>Social acceptance</i>	<i>Schooling</i>
1	Professional service	Professional degree	Rs. 1.5 lac	Putting all efforts, but all services not available.	Good	NA
2	Skilled worker	Higher secondary	Rs. 0.15 lac	Father has left. Earning is first priority for mother.	Fair	Not yet
3	Farmer	Secondary school	Rs. 0.4 lac	Parents negligent and unaware of the needs.	Fair	Not accepted
4	Farmer	Primary	Rs. 0.25 lac	Parents negligent and unaware of the needs.	Fair	Not accepted

Table 5 – Physical environment

Case No.	House	Toilet	Bedding	Streets	Conveyance
1	Pucca, ground floor	Squatting	Hard bed, firm mattress	Tar road	Two wheeler
2	Pucca, first floor	Squatting	Floor, thin mattress	Bolder road	None
3	Kuccha	Squatting	Floor, nylon rug spread on hay/twigs	Kuccha road	Two wheeler
4	Kuccha	Squatting	Jute woven bed	Kuccha road	Bicycle



Fig. 1 – Un-excised myelomeningocele sac



Fig. 2 – Multiple ulcers on buttocks and knees over last 5 years. See the rough woven bed.



Fig. 3 – Poor access to toilet due to curbs, drains and narrow doors in spite of wide spaces available

was however far away in another district. Case no. 1 had undergone surgeries for associated conditions of myelomeningocele.

The social support available to these cases (Table 4) was assessed by interviewing family members at the residence. All except case No. 1 were from lower socioeconomic strata. The medical expenditure till date varied from thousands to lacs of rupees according to the financial strength and reimbursement facilities available to the family. The social acceptance was fair in the neighborhood but the elder two cases were denied admission in nearby schools because of the bladder bowel incontinence. Parents of case no. 1 were educated, aware of the special needs and are putting their effort in appropriate direction. They expected better rehabilitation facilities in their vicinity. The family of case no. 2 was in turmoil because her father had left the family. The mother had dual responsibility of earning and taking care of the 3 siblings out of which 2 were suffering from myelomeningocele. Hence the house had poor hygiene and was full of urinary odour sufficient enough to repel an occasional visitor.

The physical environment, architecture and conveyance available to the cases (Table 5) were assessed. The first two cases had pucca house while other two had kuccha house. None of them had commode type water closet. Only case number 1 was taken to the toilet for voiding and defecation while no special effort for toilet training was made in the other three cases. Those three cases used to pass urine and stools on the bed or anywhere in the house and the excreta were cleaned by family members routinely. The parents of the elder two cases were unaware of the bladder bowel complications but were worried about the visibly obvious pressure ulcers. However they were ignorant about the high risk factors of ulceration like improper sitting/lying surface like jute (Fig. 2), rugs, poor hygiene with resultant moisture at the pressure regions and excess friction while moving them. They were not sensitized about pressure relieving techniques. The flooring had multiple levels especially at the passage to the toilets (Fig 3). The flooring was smoothed in kuccha house by cow dung finishing. The access outside the house was uneven on both tarred/untarred roads. The available conveyances for the cases

were two-wheelers and bicycle where they were carried on pillion. The case no. 2 had no conveyance available because the mother (the only caregiver) could not drive any vehicle.

Discussion

A follow up of four cases from a composite disability identification camp at Bishrampur, Sarguja, Northern Chhattisgarh are presented here to reveal the current scenario of medical, rehabilitation and social facilities available to spina bifida in rural settings visited.

The high turn out (5% of the locomotor disabled) of cases of myelomeningocele in this camp raised a query whether the condition is so highly prevalent. This camp was specifically for children below 14 year age hence congenital impairments like spina bifida may be just becoming conspicuous. This, however, may not be considered a representative sample from the population. On literature review, one of the highest incidences in world was found in remote villages of Uttar Pradesh amounting to 6.57 to 8.21 per thousand births. The incidence has shown great influence of socioeconomic status⁴, prenatal diagnostic facilities⁵ and dietary folate^{6,7,8}.

In a hospital based study in Nigeria⁹, out of 312 cases of spina bifida cystica, 90% were home delivered with no antenatal services, only one fifth reported for its treatment within 24 hours of birth and only a third underwent surgery mainly for social reasons. An important finding was that 97.5% of them were lost to follow-up after infancy. The situation may be same in India due to poor health infrastructure with virtually non-existent rehabilitation services.

The local NGO had mobilized a lot of cases for the camp from remote areas and the transport facilities were made available during the camp. The high number of spina bifida raised a curiosity to search the medical rehabilitation facilities available to these children. Hence it was decided to follow-up these cases at their residence. The children hailed from rural areas, some close to the town and some in remote areas.

The expressed need of all the cases was "walking". Good quadriceps power predicts community ambulation¹⁰ in the first two cases. The elder two cases, with high lumbar level, with extensive pressure ulcers felt that pressure ulcer needs treatment. Number and severity of the ulcers was more in the elder case. The risk of pressure ulceration was unknown to the two younger cases having a lower spinal level.

None of them posed incontinence as the prime problem even when all of them suffered from the complaint. One of the children had unbearable bad odour of urine due to dribbling but the parent was not much bothered about the

same and did not mention it as a major social problem either. This attitude of the family may hinder the development of social relations.

Schooling of the children was also a felt need of the parents irrespective of their socioeconomic strata. Out of the four children, two were in schooling age. Those two children were not accepted in local school due to poor bladder bowel control and problem of conveyance. None of the children attended school as of now.

The services available to them were appropriate diagnosis and referral for sac excision. All the children received appropriate medical diagnosis and were offered primary surgical treatment. A 4 year old case had not yet undergone sac excision for fear of complications of similar surgery in her elder brother, himself being a case of spina bifida. The surgical facilities were availed in far away districts from their residence.

Preventive measures against folate deficiency in the population are lacking. There was lack of medical follow up services for the specific condition but they had access to physician/pediatrician for general illnesses. The parents were not aware of any of the complications and necessary long term needs of these children irrespective of their socioeconomic status. The primary health care facilities were nearby except for one case where the Primary Health Centre (PHC) was 12 km away.

The awareness of rehabilitation facilities was there but the facilities of therapeutics and assistive devices were lacking. They did not have sufficient knowledge of modified clothing and appropriate diet management that may be useful in such condition.

There was a problem of conveyance and ambulation which was unsolved. This was felt more conspicuously in those with school going age. The house and street had architectural barriers posing problems of access to the cases.

The financial condition is not well enough to support the ever persisting medical requirements of the children. There is no insurance scheme to take care of their health aspects and hence the medical expenditure is to be incurred first and then reimbursement was received in one case. Thus the financial backing had lot of hassles.

Suggestions and Recommendations

It is high time that folate supplements (0.4 mg synthetic folate) be made available to the all women in India throughout their childbearing age till the family is completed. Such national efforts have given good results in terms of reducing the incidence of spina bifida in various countries¹¹.

The medical or surgical needs throughout the lifetime of the patient say for the (sac excision, hydrocephalus, limb

deformities, pressure ulcers, bladder bowel management etc.) may be made available through identification camps linked with a better referral system. The anxiety, hassles, financial hardships suffered by the cases during the referral process can be minimized by developing good network of the service providers viz. NGOs, financiers, hospitals, government agencies etc.

The long term management services can be improved by continuing training of health care professionals at tertiary as well as grass root levels. It is essential to create awareness about complications of spina bifida and its associated morbidity especially affection of the excretory system and pressure ulcers. Appropriate guidance and counseling for the same needs to be provided to the parents and the patients so that they know what problems are expected lifelong and what are the ways to prevent or overcome them.

The therapeutic inputs and fitment of assistive devices are very specialized services which can be provided through trained rehabilitation professionals in government/NGO/private sectors. However the core principle should be made clear that those services are needed throughout life. Hence the parents and the patient must get trained in their own care so that regular monthly/quarterly follow-up visits to rehabilitation facilities should suffice.

The PHCs, which are the only resource for most of the rural India, are far from being utilized of their potential to provide long term management for the condition. Their potential needs to be exploited by appropriate training of the medical personnel.

The welfare department has established apex institutes, regional centers and district disability rehabilitation centers (DDRCs) to provide rehabilitation facilities to the disabled persons. At present they are concentrating on identification and provision of assistive devices to the disabled under the ADIP scheme of the Ministry of Social Justice and Empowerment, Govt. of India. Different designs of clothing for incontinence and safety covers for hands, knees are available and can be provided under the scheme. These establishments manned by therapists, orthotists, and other rehabilitation professionals can be well utilized to provide continuing rehabilitation services to the cases.

The patient needs to be made aware of the bad odour of urine and feces and its necessary management so that he becomes presentable in society. Bladder and bowel training needs to be stressed from early childhood. Specific postures and exercises to prevent progression of deformities are essential. Independence in activities of daily living needs to be trained especially the management of toilet activity and clothing. Clean intermittent catheterization, external catheterization, diet modification, timed voiding/defecation go in a long way

to prevent severe complications and help socialization.

The Persons with Disabilities Act, 1995 comes forth to help in this regard and accordingly governments and authorities have to take up the task of barrier free access. Independent access to house, streets, playground, school, workplace and public buildings needs to be ensured by necessary modifications. The Sarva Shiksha Abhiyan comes with components of inclusion of these children into normal schools, home based teaching and conveyance facilities which is their right.

All services can be well coordinated and implemented through a parents' support group or self help group of the similar cases in the locality. One such parent support group has been formed in Mumbai in Feb. 2006¹². Such a group can become a guiding place, an emotional support and advocate for the needs of the patients as well as provide vocational and recreational avenues for the patients. General access to needful medical and surgical management can be improved by insurance and other methods of funding.

Research and development is required in the field of mobility aids, assistive devices, affordable techniques of management of hydrocephalus (low cost shunts), bladder, bowel problems, food fortification and folate supplementation.

Summary

This study reveals the scarcity of services available to spina bifida patients in rural India. Their needs of ambulation, management of bladder, bowel and pressure ulcers are largely unmet. The present infrastructure of health and welfare departments with district hospital in each district, PHC in each block, DDRC in more than 100 districts, regional centers in 10 places and the apex/tertiary level institutes can be utilized in coordination with NGO and private sectors to provide rehabilitation services to this group. There is a need to create awareness about the condition and its lifetime needs which can be met by appropriate rehabilitation management. Last but not the least, there is urgent need of instituting preventive measures against occurrence of spina bifida. This is the first task for parent group and self help groups working for the condition.

Acknowledgements

The cooperation of the patients and their family members is highly acknowledged. Dr AS Pandit, Mr Mahipal and staff of CRC-Bhopal and DDRC-Jashpur are acknowledged for help in the presentation. TEAM, Digdarshika, Bhopal and Gyanoday deaf school, Bishrampur, are specially acknowledged for their help to reach the patients.

References

1. Johnston MV, Kinsman S. Congenital anomalies of the nervous system. In: Behrman et al. 17th Edition. Nelson Textbook of Pediatrics. India. Saunders. 2004: 1983-6.
2. Gross RH et al. Early management and decision making for the treatment of myelomeningocele. *Pediatrics* 1983 Oct; 72(4): 450-58.
3. Cherian A et al. Incidence of neural tube defects in the least developed area of India: A population based study. *Lancet* 2005; 366: 930-31.
4. Nesbit DE, Ziter FA. Epidemiology of myelomeningocele in Utah. *Dev Med Child Neurol* 1979 Dec; 21(6): 754-57.
5. Luder S, Schulte FJ. Prevalence and geographic distribution of spina bifida aperta in West Germany. *Klin Pediatr* 1989 Mar-Apr; 201(2): 73-77.
6. Medical Research Council Vitamin Study Research Group. 1991. Prevention of neural tube defects: results of the Medical Research Council Vitamin Study. MRC Vitamin Study Research Group. *Lancet*; 338: 131-37.
7. Cziezel AE, Dudas L. Prevention of the first occurrence of neural tube defects by periconceptional vitamin supplementation. *N Engl J Med* 1992; 327: 1832-35.
8. Berry RJ et al. Prevention of neural tube defects with folic acid in China. China-US collaborative project for neural tube defect prevention. *N Engl J Med* 1999; 341: 1485-90.
9. Mabogunje OA. Spina bifida cystica in northern Nigeria. *Childs Nerv Syst* 1990 Mar; 6(2): 103-6.
10. Selber P, Dias L. Sacral level myelomeningocele: long term outcomes in adults. *J Pediatr Orthop* 1998 Jul-Aug; 18(4): 421-22.
11. Freire WB et al. Effect of folic acid fortification in Chile: preliminary results. *Eur J Pediatr Surg* 2000; 10(Suppl 1): 42-43.
12. Inauguration of Parent Support Group *IN* 1st National Update on Spina Bifida & Hydrocephalus *AT* Tata Memorial Hospital, Mumbai on 18th Feb 2006. www.spinabifidaclinic.in