ORIGINAL ARTICLE

Hydrocephalus in Children with Spina Bifida

Sanjay Sadhu, H.L. Goswamy, S. K. Gupta*, Inder Singh**,
Nasib C. Digra, Ravinder K. Gupta***

Abstract

A prospective study consisting of 50 infants having spina bifida and cranium bifidum and relationship of hydrocephalus was carried out. The male preponderance was obvious (1.7:1) with majority of infants being less than 3 months. The presenting complaint in almost all the children was swelling on back or head. Lumbar spine was affected most commonly with 36% children having lesion at this site followed by lumbosacral (20%) and occiptal (16%). Neuromuscular deficit was present in 32% cases. Clinical evidence of hydrocephalus was seen in 22% children. X-ray skull was suggestive of hydrocephalus in 22% cases. Ultrasound examination of skull confirmed the diagnosis in 52% cases. Hydrocephalus had no specific relationship with site of lesion. It was more commonly associated with neurological deficit. Sonographic examination is relatively cheaper and invaluable investigation, and should be done routinely in patients with spina bifida.

Key words

Spina bifida, Cranium bifidum, Hydrocephalus

Introduction

Spina bifida is one of the commonest congenital, neurological malformations in children. Its incidence varies from 0.69 per 1000 births to 2.53 per 1000.(1-4) Inspite of lot of progress, the outlook for children with spina bifida is not always good. Many of them also have associated abnormalities like hydrocephalus, paraplegia, sphincteric incontinence and deformities of the lower limbs which are responsible for the morbidity and mortality in these children. Hydrocephalus is present in a large number of children with spina bifida mostly because of associated Arnold-Chiari malformation but can also be because of factors like deformities of aqueduct.(5-7) In many children hydrocephalus becomes evident for the first time after surgery for spina bifida. Surgery can sometimes aggravate hydrocephalus and some patients may have rapid progress of hydrocephalus in the immediate post-operative period requiring urgent intervention.(8,9) Hydrocephalus in addition to affecting the final outcome of these children, is also a major factor like neurological deficit in deciding the course of treatment of a child with spina bifida. The recognition of

From the Postgraduate Departments of Surgery, *Medicine, **Radiology & ***Paediatrics, Govt. Medical College, Jammu (J&K).
Correspondence to : Dr. Sanjay Sadhu, 9-A, New University Campus, Jammu (J&K) India.

hydrocephalus thus is important for deciding the management of these children. The study was undertaken to see association of hydrocephalus with spina bifida in children.

Material and Methods

The study was conducted on fifty children of spina bifida and cranium bifidum. After obtaining history and doing detailed general physical as well as neurological examination, these patients were subjected to roentgenography of skull and ultrasound examination. Ultrasound examination was done by using machine Sanoline-2 manufactured by Siemens and RT 3000 manufactured by General Electric. It was done by using sector probe of 3.5 MHz and 5 MHz. Scans were performed in coronal, sagittal and axial planes. Anterior fontanella was used as window for obtaining coronal and sagittal views.. Also CT scan was done in 15 children, who could afford it.

Results

Out of the fifty children studied, majority (58%) were neonates with a definite male preponderance (62%). The presenting complaint in almost all the children was swelling on back over the spine or at the back of head. Ten children (20%) had weakness of lower limbs and 3 (6%) had complaints pointing towards hydrocephalus. First born were affected more frequently (52%). There was no history of similar abnormality in the family in any case. Lumbar spine was affected most commonly with 18 (36%) children having lesion at this site, followed by lumbosacral (20%) and occiptal (16%). Two children in the study had double lesion, one child had the lesion at occiptal and lumbosacral regions and the other one at cervical and lumbar spine, as shown in Table I. Neurodeficit was present in 16 (32%) chlidren with 7 children having paraparesis and 9 having paraplegia. Four children had asymmetrical neurodeficit. Hydrocephalus was seen to be more frequently associated with spina bifida when the child also had neuromuscular deficit. Out of 16 children who had neuromuscular involvement, 14 (87%) had associated hydrocephalus as shown in Table II. Sphincteric involvement was observed in 28% cases. Eight (16%) had other associated congenital abnormalities. Clinical evidence of hydrocephalus in the form of increased head circumference, tense fontanelle, widening of the sutures and sunset sign was observed in 22% children. X-ray skull was suggestive of hydrocephalus in 22% cases. Ultrasound examination of the head confirmed the diagnosis of hydrocephalus in 26 patients giving an incidence of hydrocephalus in spina bifida of 52%. CT scan done in 15 such cases also confirmed hydrocephalus.

Table 1. Site of lesion of spina bifida and hydrocephalus.

Site of lesion	No.	%age	Hydrocephalus Absent	Hydrocephalus Present	Hydrocephalus %age
Occipital	8	16	3	5	62
Cervical	2	4	2	—	—
Dorsal	6	12	3	3	50
Dorsolumbar	2	4	—	2	100
Lumbar	18	36	8	10	55
Lumbosacral	10	20	5	5	50
Sacral	2	4	2	—	—
Double lesion	2	4	1	1	50
Total	50	100	24	26	52

Table II. Neurodeficit and hydrocephalus in children with spina bifida.

	No.	Hydrocephalus Absent	Hydrocephalus Present	Hydrocephalus (in %age)
Children without neurodeficit	34	22	12	35
Children with neurodeficit	16	2	14	87
Total	50	24	26	52

Discussion

The incidence of spina bifida varies from 0.69 per 1000 births to 2.53 per 1000 births.(1-4) A higher incidence upto 11.4 per 1000 births have been reported in Karnataka.(4) In addition to varying degrees of neurodeficit hydrocephalus is the commonest associated abnormality obsreved in these children. It occurs because of associated Arnold-Chiari malformation in these children.(10) Some of the children also have aqueductal abnormalities which can result into hydrocephalus.(11)

Males (62%) predominated in our study. Most of the studies have reported spina bifida to be commoner in females.(1,2) It is possible that social bias against females in the Indian society is the cause of males outnumbering females. Although no definite pattern of inheritance has been observed in spina bifida, it has been seen that first born children are most commonly affected.(12) The distribution of site of lesion in our study is similar to that observed by other workers.(11,12) The commonest site in our study was lumbar spine as shown in Table I. As mentioned in literature the lumbosacral region accounts for atleast 75% of the cases.(13) Varying degrees of weakness of lower limbs were seen in 16 (32%) of our patients with 7 patients having paraparesis and 9 patients having paraplegia. The incidence is less than those reported by other workers.(11,14) It has been mentioned that neurological deficit is maximum with thoracic and high lumbar lesions. A variety of problems are encountered i.e., variable degrees of lower motor neuron paralysis of the lower extremities, sensory loss in the perineal area and lower limbs, sphincter disturbance and neurogenic bladder and bowel.(15) Hydrocephalus clinically suspected in 22% patients was much lower than reported by others.(8,16) Sonographic examination of head revealed 52% hydrocephalus as compared to 95% to 96.5% as reported in different studies.(6,17) The lower incidence of hydrocephalus in our study may be because of small number of patients. The incidence of hydrocephalus also varied according to the site of the lesion. Out of the 8 patients of occiptal encephalocele, 5 (62.5%) had hydrocephalus. None of the two children with cervical lesion had hydrocephalus. The incidence of hydrocephalus in patients of dorsal spina bifida was 50% with three out of six having hydrocephalus. Both the children with dorsolumbar spina bifida in this series had hydrocephalus giving an incidence of 100% in them as shown in Table I. One patient out of the two with double lesions had hydrocephalus. Thus in our experience it was most commonly associated with lesions in dorsolumbar region and in cases of occiptal encephalocele in contrast to other regions reported by other workers.(11, 18) It has been mentioned in literature that generally the lower the deformity in the neuraxis (e.g., sacrum), the less likely is the risk of hydrocephalus (13). It was a contrast finding in our series.

Out of 26 patients with hydrocephalus on ultrasound examination, 15 (58%) did not manifest it clinically as has been observed by various other workers following pneumoventriculography.(12,18) Out of 39 patients having normal head circumference 15 (38%) had hydrocephalus as compared to 69% reported in one of the series, probably because of higher incidence of patients having hydrocephalus (18).

In our study hydrocephalus was seen to be more frequently associated with spina bifida when the patient also had neurodeficit. Out of 16 children who had neuromuscular involvement, 14 (87%) had associated hydrocephalus. The clinical examination was found to be inaccurate in detecting hydrocephalus and only 42% children having hydrocephalus on sonographic examination manifested clinically.

Hydrocephalus was present in most cases of spina bifida. It has no specific relationship with the site of lesion. It was more commonly associated with neurological deficit. Sonographic examination is relatively cheaper and valuable investigation and

should be done routinely in patients with spina bifida and cranium bifidum.

References

1. MacMohan B, Pugh TF, Ingalla, TH. Anencephalus, spina bifida and hydrocephalus. Incidence related to sex, race and season of birth and incidence in siblings. *Br J Prev Soc Med* 1953; 7 : 211-19.
2. Wiswell, TE *et al.* Major congenital neurological malformations. *Am J Dis Child* 1990; 144 : 61-67.
3. Nash, DFE. Meningomyelocele. *Proc Roy Soc Med* 1963; 56 : 506
4. Kulkarni ML, Mathew MA, Ramachandran B. High incidence of neural tube defects in South India. *Lancet* 1987; 1 : 1260.
5. Babcock DS. Sonography of congenital malformationis of the brain. *Neuroradiology* 1986; 28 : 428-39.
6. Babcock DS, Han BK. Cranial sonographic findings in meningomyelocele. *Am J Roentgenology* 1981;136:563-69.
7. Babcock DS, Han BK. The accuracy of high reslution, Real-time ultrasonography of the head in infancy. *Radiology* 1981; 139 : 665-76.
8. Doran PA, Guthkelch AN. Studies in spina bifida cystica. I. General survey and reassessment of the problem. *J Neurol Neurosurg Psychiat* 1961; 24 : 331-45.
9. Gorezyca JT, Banta JV. Vertebral anomalies associated with spinal dysraphism. *Eur J Paed Surg* 1993; 3(suppl 1) :27.
10. Ingraham FD, Scott HW. Spina bifida and cranium bifidum. V. The Arnold-Chiari malformation: A study of 20 cases. *New Eng J Med* 1943; 229 : 108-14.
11. Laurence KM. The natural history of spina bifida cystica. *Arch Dis Child* 1964; 39 : 41-57.
12. Lorber J, De NC. Family history of congenital hydrocephalus. *Develop Med Child Neurol* 1970; 12 (Suppl 22) : 94-100.
13. Haslam RHA. Congenital abnormalities of the central nervous system. In : Behrman RC, Kliegman RM, Jenson JB (eds), Nelson Textbook of Pediatrics, 16th edn. WB Saunders Company, Philadelphia, 2000 : 1803-05.
14. Kalra V, Kumar R. Normal development and malformations of central nervous system. In : Parthasarthy A, Menon PSN, Nair MKC (eds) IAP Textbook of Pediatrics, 2nd edn. Jaypee Brothers, New Delhi, 2002 : 294-95.
15. Upadhyaya P, Singh N, Parthasarthy V. Cystometric evaluation of the functional status of neurogenic bladder in children with meningomyelocele. *J Paed Surg* 1974; 9 : 477-83.
16. Eckstein HB and MacNab GH. Myelomeningocele and hydrocephalus. The impact of modern treatment. *Lancet* 1966 (i) : 842-845.
17. Jayakrishnan VK *et al.* Cranial sonographic findings in patients with meningomyelocele. *Ind J Radiol Imag.*, 1993; 3 : 9-13.
18. Lorber J. Systematic ventriculographic studies in infants born with meningomyelocele and encephalocele. *Arch Dis Child* 1961; 36 : 381-89.