

# The spectrum and challenges of spinal disorders presenting to a pediatric Orthopedic Hospital and Rehabilitation Center

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The management of pediatric patients with spinal disorder poses a significant challenge to the treating physician in the context of limited resources and unlimited number of patients. Successful management of these cases involves a dedicated team of orthopedic surgeons, rehabilitation physicians and nurses apart from a highly motivated family and friends. The dilemma of undertaking major intervention versus permitting a child to deteriorate to a worsening level of function and life quality is indeed painful and distressing. This study reviews the challenges we are facing in the management of spinal disorders at our tertiary level center.

**Key Words:** late presentation, spinal deformity in children, retrospective study, tuberculosis

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The Pediatric Spine poses unique challenges to the treating orthopedic surgeon and these challenges are compounded many folds when the patient presents late for treatment. Thus it is frequent to encounter many spinal disorders at the Hospital and Rehabilitation Center for Disabled Children (HRDC) (in the Hospital OPD and Mobile Camps) which are either untreatable or pose immense risks of serious complications with intervention. The dilemma of undertaking major intervention versus permitting a child to deteriorate to a worsening level of function and life quality is indeed painful and distressing. In this retrospective study we review our experience in the management of spinal disorders at our center.

## Materials and Methods

The HRDC is a tertiary level hospital situated about 25 kilometers from Kathmandu and caters for all kinds of pediatric cases including orthopedic and spinal diseases. The hospital records of patients with spinal disorders

registered with HRDC from 1995 through 2001 were retrospectively reviewed. One hundred and sixty-five children less than 17 years of age with structural lesion in the spine, with or without neurological deficit, were admitted during the period under review and these patients constitute the basis of this study. Details of the patient's presenting symptoms, neurological deficits, and physical deformity were recorded. Management of the patient according to the diagnosis established at our center was recorded and short-term outcome at discharge from the hospital analyzed.

## Results

A total of 165 patients were reviewed during this period and three main categories of disorders were attributable, namely tubercular infection, trauma and congenital deformities.

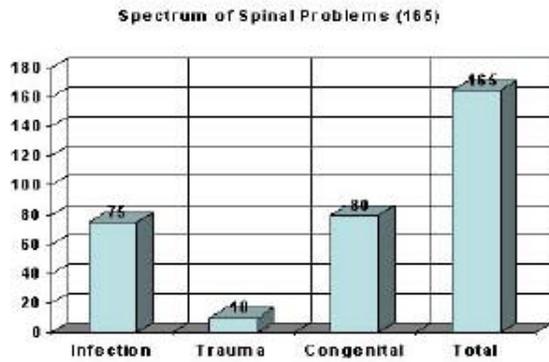


Figure 1. Bar chart showing the spectrum of spinal diseases at HRDC

### Tubercular spondylitis

Tubercular spondylitis accounted for 75 (45.4%) of the total number of cases. Age ranged from 3 years to 16 years with the average being 11.7 years. Male and female children were almost equally affected and the pattern of involvement was not unusual. Only 17 cases (22.7%) (male 10, female 7) had neurological deficits, duration ranging from 15 days to 18 months. Sixteen cases were paraplegic and one case that had involvement of the cervical spine was quadriplegic. The site of involvement of the spine by tuberculosis in all patients is shown in Figure 2 and those with neurological deficits is depicted in Figure 3.

All the 17 cases underwent surgical decompression. Neurological recovery was observed in all cases. One case had relapse and re-decompression was performed. However the patient failed to show any improvement. One patient died of re-exacerbation with abscess extension into the mediastinum, 9 months after the initial recovery.

All the remaining 58 (77.3%) cases presented with kyphotic deformity with minimum kyphotic angle measuring 30 degrees to maximum of 90 degrees (average 69.6 degrees). There were 56 patients in this series with gibbus angle measuring over 60 degrees. There were 11 patients in this group that presented to us with severe deformities and compromised pulmonary function that we labeled "untreatable." Figure 4 shows the typical xray of the patient with tubercular spine with a kyphotic deformity. Some patients with neurological deficit and kyphotic deformity had associated contractures of the hip and knee, which were intensively treated with traction and physical therapy, in addition to definitive treatment (Figure 5).

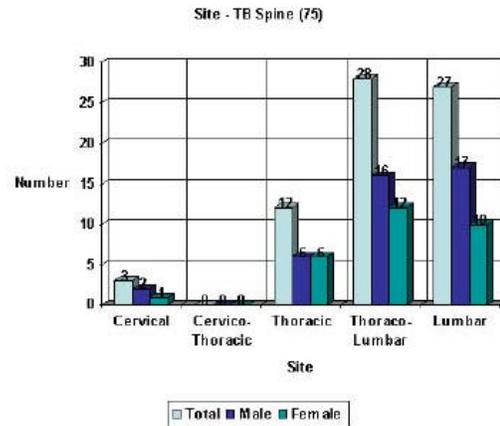


Figure 2. Bar chart showing the anatomic level of involvement in the tubercular spine patients

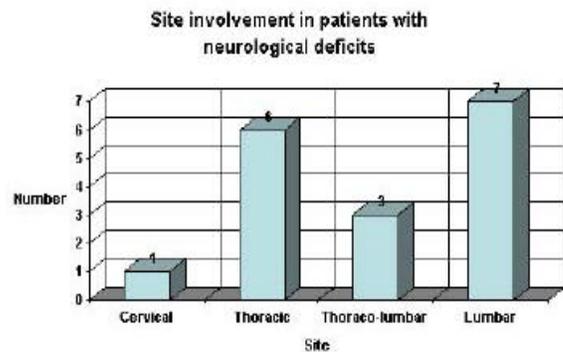


Figure 3. Bar chart showing the site of involvement of 17 patients with tubercular spine with neurological deficit.

All the cases with tuberculous spondylitis were treated with 12 to 18 months of anti-tubercular chemotherapy consisting of the standard 4 drugs regime (Isoniazid, Rifampicin, Ethambutol and Pyrizinamide) for 3 months and 2 drugs regime (Isoniazid, Rifampicin) for the remaining months.

This category of patients fared best of all. Even those patients with moribund condition responded well to a combination of aggressive nursing care, chemotherapy and surgery (anterior decompression and fusion). Recumbency was emphasized in all patients and protracted spinal orthotic support was used to prevent worsening of spinal deformity. The pediatric spine is largely cartilaginous and achieving a fusion mass is challenging, in those patients who are operated. Recovery from neurological deficit after

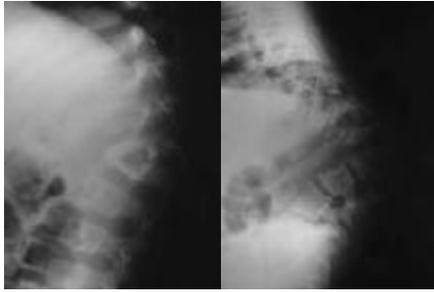


Figure 4: Plain X-rays lateral views showing vertebral destruction with anterior wedge compression leading to severe kyphotic deformity.

surgical decompression was observed as early as 7 days. In our series all patients recovered to full motor power.

#### Trauma

There were only 10 cases admitted for evaluation and management of major complications following spine trauma. As shown in Figure 6 the thoracic spine was the most frequent site of injury, followed by the lumbar region.



Figure 5. Seven years old boy (left) and his x-ray of lumbosacral spine (center and right), a case of TB Spondylitis of Lumbo-Sacral region, presented with severe Hip/Knee contracture. Patient being treated with chemotherapy. Skeletal traction is applied to correct Hip/Knee contracture.

The cases were all referred from other hospitals and presented with severe functional difficulties. All the cases were paraplegic with bowel/bladder incontinence. Duration at presentation to our center ranged from 15 days to 10 months. Figure 7 shows an example of patients with traumatic paraplegia after intensive rehabilitation.

Seven patients (70%) had large decubitus ulcers at trochanteric and sacral areas. Three patients underwent gluteal flap rotation to cover the defects. Other cases were treated with regular change of posture and frequent dressings changes. None of these patients underwent any surgical treatment. Our treatment consisted of pressure sores care and rehabilitation aimed at maximal functional gain. None of the patients were ambulatory ( we do not

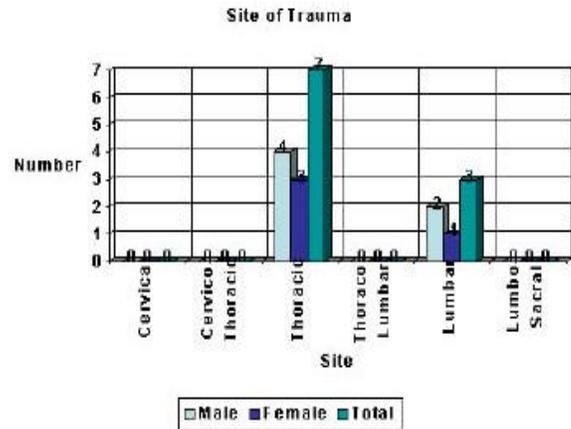


Figure 6. Pattern of involvement of spine following trauma

receive acute trauma at our center, and the cases seen here were all referred from other centers). The focus of our management was restoring maximal function both in terms of activities of daily living (ADL), bowel/bladder care, skin care and locomotion whenever this was feasible.



Figure 7. Thirteen years old girl fell from a height 2 years back. She presented with paraplegia and Bowel/Bladder incontinence one month after the injury. No neurological improvement has been observed thus far. She underwent intensive rehabilitation and was independent for ADLs on a wheelchair at time of discharge.

#### Congenital spinal deformities

There were a total of 80 patients in this category, including 38 (47.5%) cases of spina bifida, 32 (40%) cases of scoliosis and 10 (12.5%) cases of congenital kyphosis (Figure 8).

There were 38 cases of spina bifida. Twenty nine (76.4%) cases were spina bifida occulta whereas 9 (23.6%) cases were with cystica. Figure 9 shows an otherwise

uncomplicated case of myelomeningocele whereas

**Congenital spinal deformities (80)**

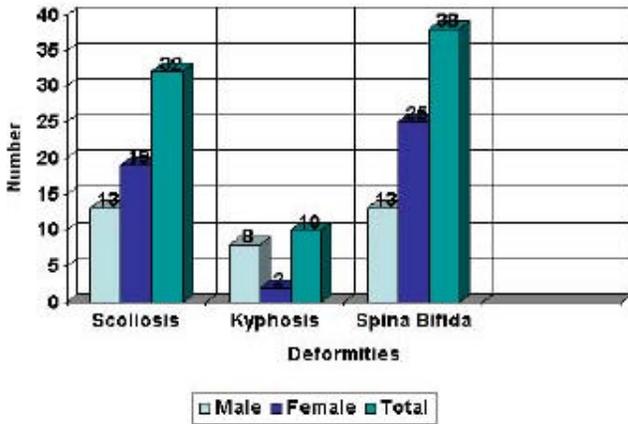


Figure 8. Pattern of congenital spinal deformity at HRDC



Figure 9. Seven years old boy with Myelomeningocele. Except for sensory loss over the lateral aspect of the foot, he has no other neurological deficits



Figure 10. Thirteen years old girl with Myelomeningocele at Thoraco-lumbar region, presented with severe contracture of Hip/Knee and huge bed sore at the gluteal region. Bed sore has been present for the last 3 years

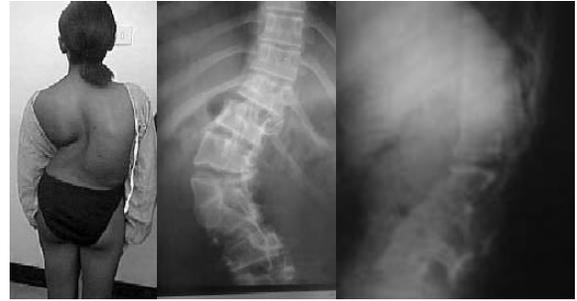


Figure 11. Thirteen years old girl with Congenital Scoliosis (right) at Thoraco-Lumbar region. Xrays (right and center) showing formation and segmentation defect of L1, L2 vertebral bodies.

Figure 10 shows a girl with myelomeningocele with decubitous ulcer managed at our center.

It is to be noted that in most cases primary repair of the spinal defects was done elsewhere by either neurosurgeons or pediatric surgeons. All cases of spina bifida occulta presented with foot deformities. Evaluation of the back revealed a patch of hair or a skin dimple. Radiological evaluation revealed bony defect or deformity in the posterior vertebral arches.

Among the 38 cases of spina bifida, 29 cases (76.4%) were found to have treatable pathologies like clubfoot, Vertical Talus and Pes calcaneus deformities. Seven patients (18.4%) had non-healing trophic ulcers in the foot. Four patients with severe infection underwent below knee amputation (one had bilateral below knee amputation). Another 3 patients are being treated with total contact cast and modified shoes with soft cushion. If these ulcers are taken care of properly and adequately, total contact cast and shoe modification with soft cushion seemed to be an effective treatment.

**Kyphosis (10)**

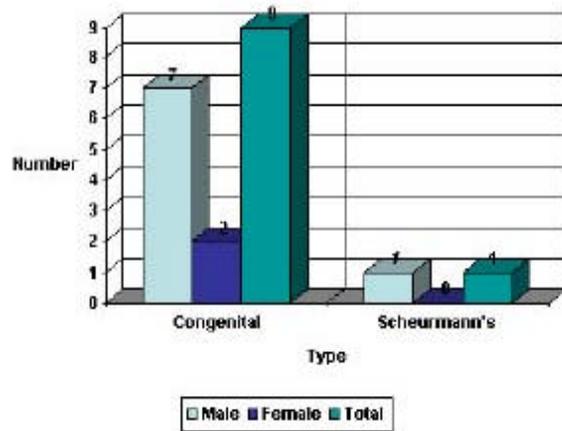


Figure 12. Bar chart showing the type of kyphosis.

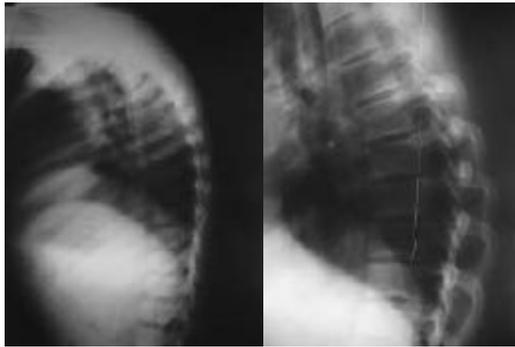


Figure 13. X-rays of Thoracic spine lateral views showing Round Back Deformity

Of the 32 cases of scoliosis, congenital scoliosis is seen more frequently at our hospital as compared to idiopathic scoliosis (21 vs. 11). The Cobb angle ranged from 20 degrees to 90 degrees, the average being 43 degrees. Formation defect was the most common pattern of congenital deformation. Figure 12 shows an example of a case of congenital scoliosis.

There were 10 cases of Spinal Kyphosis (Figure 12), (90%) being congenital and 1 (10%) due to Scheurmann's disease. Six cases had Round Back Deformity (Figure 13), and three had formation defect of vertebral bodies.

Scoliosis and Kyphosis were largely severe deformities with gross uncosmesis, this being the major concern of the guardian. The majority of these patients were "untreatable". Five patients with scoliosis underwent surgery (fusion) aimed at stabilization of the curve in situ. In house training of surgeons and acquisition of equipment for safe instrumentation of the spine is an ongoing project at our center at present. All other cases were braced.

### Discussion

Nepalese patients have limited access to specialized medical care and our experience at HRDC with patients presenting to us with spinal disorders is unique. The majority of cases that have visited our center over the past 7 years have severe deformities or very significant complications which have either been unattended or inadequately so.

Tubercular spondylitis continues to challenge us. However, we would have however expected to encounter a larger number of patients with tuberculosis of the spine, given the wide prevalence of tubercular infection in Nepal. One of the reasons may be the tertiary nature of our center receiving only a special category of complex cases. The role of anterior decompression in tubercular spondylitis is well established,<sup>2,6</sup> and our experience bears this out. The

challenge is to achieve a fusion mass and prevent long term spinal deformity. External bracing has an important role and this needs to be emphasized to lessen the severity in the long run of the gibbus which is seen in a large proportion of cases in our series. Early recognition and initiation of anti-tubercular chemotherapy and judicious surgery and bracing are all important components in the management. Despite the difficulties posed by patients presenting primarily for the treatment of severe deformities, those patients with neurological deficits and abscess formation respond well to treatment.

All the 10 patients with spine trauma were old cases with serious complications. All the children were incontinent and 7 patients had large pressure sores. The children were in poor states of nutrition and had lost self confidence and esteem. The vibrant positive environment at HRDC made a very big difference right from the very outset. With diligent nursing care and rehabilitation assistance we were successful in healing all the decubiti. Three patients required a flap rotation to heal a very large trochanteric decubitus ulcer. Spinal trauma in the pediatric age group is less common as compared with the adult,<sup>4</sup> but the consequences of major neurological catastrophe in a child can be the ultimate challenge. For better management of these challenging cases a closer liaison between orthopedic surgeons, neurosurgeons, rehabilitation specialists and physiotherapists is crucial. Our goals at HRDC are focused in this direction but there are many shortcomings. Programs focused on public education and prevention of spinal trauma need to be implemented. It is very disconcerting to have children with severe spinal deformities or spinal disorders with catastrophic sequelae presenting to our center late for treatment. Because a large number of patients with scoliosis and kyphosis were considered "untreatable", we have instituted a school screening program in Kavre district and Kathmandu to see if early detection was feasible. The role of screening programs in scoliosis is not without controversy.<sup>7</sup> We are encouraged by the screening study reports from Sweden<sup>3</sup> which indicated improved clinic attendance by patients with lesser severe deformities. At the same time, the need to further develop our surgical expertise for safe spinal instrumentation, suited to our need and context, is an ongoing exercise at our center. The orthotic/prosthetics workshop at our hospital conducts ongoing clinical trials with a variety of materials for improved bracing for spinal deformities.

Our services for patients with spinal dysraphism need to be significantly augmented both for accuracy of diagnosis and timely intervention. Children presenting with foot and ankle deformity need to be screened for possible spinal deformities. The high incidence of equinus deformity, calcaneus deformity and clubfoot in association with spina bifida is well documented.<sup>1</sup> Three important factors identified with better long term results in these patients are early expert closure of defects (in patients with midline defects), prevention of hydrocephalus and the promotion of a sitting posture so that the hands are free to interact in the

environment.<sup>5</sup> We are hopeful that we will be able to address these issues more correctly in the years ahead.

### Conclusions

Spinal disorders in children are challenging problems and much work is yet to be done in our country focusing on the problems outlined. We have made a small beginning at HRDC and there are more lessons to be learnt.

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