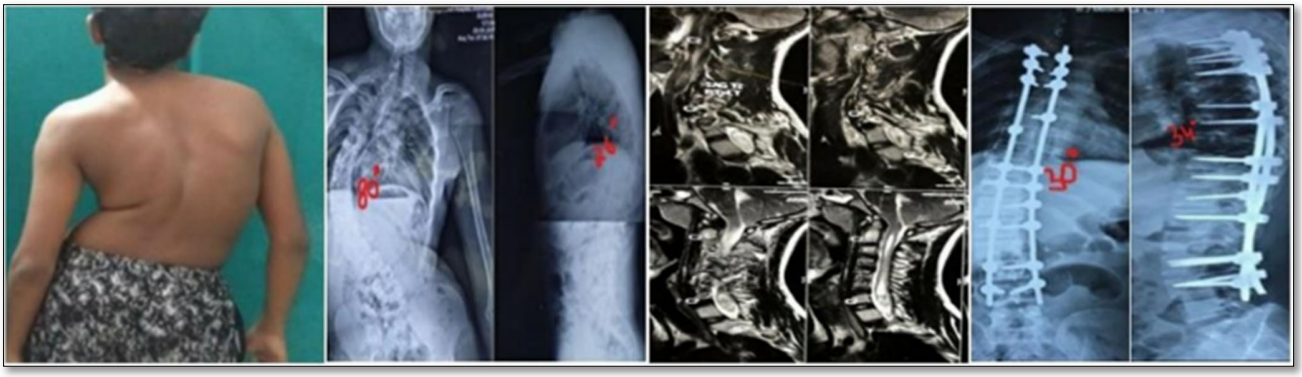

Figure 3. 16-year male with congenital right sided kyphoscolitic deformity with normal neurology which was operated for Arnold Chiari malformation and cerebella tensile herniation with extensive syrinx and spina bifida occulta at C7 -D1 level; preoperative standing AP and lateral radiograph with cob's angle 80º and hypnotic angle 26º; immediate post-operative standing AP and lateral radiograph with correction of deformity and cob's angle is 30º and hypnotic angle 34º.

DISCUSSION

The kyphoscoliosis is related with congenital anomalies in other systems, especially those formed from mesenchymal tissue. These anomalies are often asymptomatic and absent or asymmetric abdominal reflex may remain isolated finding in examination and may remain undetected until the patient is fully assessed following the diagnosis of congenital spinal deformity [4,5].

Theoretically, intraspinal abnormality can predispose the patients to neurological injury when they undergo surgical correction of scoliosis due to excessive traction of spinal cord. However, performing surgery for the intraspinal in asymptomatic patients also carries some risk of morbidity including neurological deterioration and wound problems. However, the neurosurgical procedure performed by laminectomy over 4-5 levels and flavectomy over these levels provide some form of posterior releases around the apex so. In our series of 20 patients, balancing of the S- shaped curves was achieved, and none of the patients experienced deterioration in their neurologic status after surgery.

Rapid surgical correction of severe scoliosis associated with forceful intra-operative maneuvers can increase the risk of neurologic compromise especially in patients with a history of intraspinal anomaly and we do not advocate, since the goals of the surgery are balancing of the curve, preventing progression of deformity or development of neurodeficit. Dysplastic pedicles must be anticipated in the pre-operative planning and get actual axial cut across the pedicles in CT scan and if found dysplastic, sub laminar wires, hooks and cables must be in the backup.

According to Hamzauglu [2] simultaneous surgical treatment for congenital deformity and intraspinal abnormality does not involve significant complications and seems to be an attractive and safe treatment option. It does not increase the surgical time, blood loss, and hospitalization time too much.

According to Winter [6] the classic advocated approach in patients having congenital spine deformity associated with intraspinal abnormality is first to perform surgery for the intraspinal pathologies and then surgery for correction and stabilization of the deformity 3 to 6 months later in a different session. Our study also in favor of two staged correction surgery this allow decrease morbidity of patient post operatively and allow them to recover.

Average age at which a patient presented and female preponderance of congenital scoliosis and intraspinal anomalies and thoracolumbar curve was the most common deformity in our study all was similar to all the previous reports or McMaster series [7,8].

Genitourinary anomalies that can cause obstruction are silent and, if not searched for, may cause serious damage before they are diagnosed. The incidence of genitourinary anomalies was found to be 23.8% in the current study. This is similar to frequencies seen in other studies also [8,9].

Cardiovascular anomalies were the most common of the associated systemic anomalies in our study. They were seen in 28.5% of patients with mitral valve prolapse being the commonest anomaly. According to Winter, the incidence of congenital heart disease in congenital scoliosis was 7% [9,10]. The cause of our high rate may be the small size of the patient group.

Patients with rigid spinal deformity often combine with severe pulmonary function impairment, and many research have shown that anterior spinal surgery had great impact on pulmonary function. PVCR has first been introduced by Suk et al and has been popularized by Lenke [11,12] for severe deformities of the spinal column, it enables translational and rotational correction of spinal column and offers the advantage of a controlled manipulation of both the anterior column and the posterior column at the same time through a single approach. PVCR could not only correct the deformities as good as or better than other surgery, but also

could avoid anterior approach related complications, particularly to pulmonary function impairment.

One major concern with PVCR technique is the potential neurologic complications, which may occur from direct neurologic injury during bone resection or deformity correction. The former may be mitigated through techniques such as careful retraction of nerve roots during resection of pedicles and careful planning of osteotome trajectory and force when removing the vertebral body. Neurologic complications may also result from subluxation of the spinal column, dural buckling and compression of the spinal cord by residual bone and soft tissues in the canal after correction [13].

Intraspinal anomalies were associated with 47% of patients with congenital scoliosis in the Indian population. 71% of congenital scoliosis patients with intraspinal anomalies were females. Hemivertebrae was the most common vertebral anomaly in scoliosis, and it was most located in the thoracic spine. Intraspinal anomaly was most common in vertebrae with segmentation and mixed defects. Tethered cord was the most common intraspinal anomaly in the Indian population. Special attention should be given to physical examination and inspecting for any neurocutaneous markers; however, absence of neurocutaneous markers does not rule out intraspinal anomaly. MRI should be the part of routine protocol for assessment of congenital scoliosis patients [1-3,8,12].

In the current study, the incidence of intraspinal anomalies was 35%, which was within the range reported to vary from 18.3% to 47%. Tethered cord syndrome due to tight filum terminal was the most common type of spinal anomaly noted in this study which was similar to observations reported in other studies. The mean age of onset of deformity was 11.4 years in those who had intraspinal anomalies as compared to the AIS group which was 13.2 years indicating that early age of onset is an indicator for presence of such anomalies? There is a significant correlation between age of onset of deformity and intraspinal anomaly in the idiopathic group of patients ($P \leq 0.05$). In the current study, all the patients with intraspinal anomalies associated with congenital scoliosis needed neurosurgical intervention before surgical correction of the deformity.

CONCLUSION

The Two staged surgical treatment for congenital deformity with intraspinal abnormality does not involve significant complications and seems to be an alternative and safe treatment option with respect to reduced operative time and less blood loss as compared to single stage procedure and proves to be a safe management principle to such challenging problem.

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