INTRODUCTION

The term Congenital Kyphoscoliosis describes an abnormal curvature of the spine in both coronal and sagittal plane, is a spinal deformity that results from an embryonic insult of the bony spinal column at around 5th week of intrauterine life. It is classified by the type of developmental abnormality into either a failure of formation or a failure of segmentation, however a few instances may result from a combination of both. Incidence of vertebral anomalies has been estimated to be 0.05-0.1% of live births (1). Several theories exist as to the etiology of congenital scoliosis. The overall impression is that cause is multifactorial (2-4). The developmental insult is typically not limited to the spine alone, therefore child needs to be screened for associated anomalies within and outside the spine. The primary goal of treatment of congenital scoliosis is to prevent or treat kyphotic deformities, avoid neurological deterioration, while maximizing spinal growth to the extent possible and improve the quality of life.

DISCUSSION

Knowledge about the natural history of these congenital deformities is essential as it dictates the prognosis and treatment. A large study by McMaster and Ohtsuka found that only 11% of cases were nonprogressive, 14% were slightly progressive and the remaining 75% were significantly progressive (5). Any child with short stature and spine anomalies need a detailed history, physical examination, skeletal survey with an

Case Report

We report a case of a 2 and half year old developmentally normal child born of 3<sup>rd</sup>degree consanguineous marriage who presented with abnormality of the spine since birth, short neck associated with decreased range of motion. On examination child was wasted and stunted with both weight and height falling below the 3<sup>rd</sup> percentile. Physical examination revealed a tuft of hair at the mid dorsal level with Kyphoscoliosis and right limb shortening. Kochs workup was negative. MRI Spine was suggestive of severe Kypho-Scoliotic deformity of the cervico dorsal spine. After meticulous screening the child has been referred to pediatric orthopedic department for corrective measures.

ABSTRACT

The term Congenital Kyphoscoliosis describes an abnormal curvature of the spine in both coronal and sagittal plane, is a spinal deformity that results from an embryonic insult of the bony spinal column at around 5<sup>th</sup> week of intrauterine life. The developmental insult is typically not limited to the spine alone, therefore child needs to be screened for associated anomalies within and outside the spine.

We report a case of a 2 and half year old who presented with abnormality of the spine since birth. Physical examination revealed a tuft of hair at the mid dorsal level with Kyphoscoliosis and right limb shortening. Adetailed neurological examination was performed which was unremarkable. There were no associated organ system anomalies. MRI Spine was suggestive of severe Kypho-Scoliotic deformity of the cervico dorsal spine. After meticulous screening the child has been referred to pediatric orthopedic department for corrective measures.

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MRI and other radiographic evaluation. The physical examination must not only explore the spinal deformity but also focus on chest, foot deformities and cutaneous lesions especially café-au-lait spots, sacral dimple, hair patch overlying the spine or skin tag along with genital examination.

A detailed neurological examination should be performed to rule out any associated neurologic diagnosis. Child needs to be screened for other organ system anomalies. Nearly 60% of patients with congenital scoliosis have other developmental malformations. Genitourinary abnormalities are identified in 20-40% of these children which include unilateral renal agenesis, ureteral duplication, horseshoe kidney, and genit al anomalies. Letts et al found that 82% of patients with congenital scoliosis had associated malformation in four different organ systems. Highest on the list were anomalies of the genitourinary tract. Research conducted by Mac Ewen et al revealed a 20% incidence of urinary tract anomalies. Therefore renal ultrasonography and MRI are mandatory to accurately diagnose renal anomalies.

A careful cardiac examination and echocardiography is of great concern in these children as many as 10-15% children have been noted to have congenital heart defects.

Spinal dysraphism is seen 15-40% of these children therefore a spinal MRI is indicated when there is suspicion of an underlying spinal cord abnormality.

If the diagnosis is made early, while the curvature is still small, an opportunity exists for prophylactic surgery to balance the growth of the spine. While parents are primarily concerned about the resulting cosmetic abnormalities, the physician must consider both the potential for underlying causes requiring treatment and the patients long term prognosis. The primary goal of treatment of congenital scoliosis is to prevent or treat kyphotic deformities, avoid neurological deterioration, while maximizing spinal growth to the extent possible and improve the quality of life.
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