A Genesis of Sacral Vertebrae
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Abstract: 8 years old male patient came to the hospital unable to stand and move. His lower limbs are weak and there is a small degree of deformity.

Keywords: CRS, X-ray.

INTRODUCTION
Caudal regression Syndrome (CRS) also known as Caudal Dysplasia and Sacral Agenesis Syndrome, is an uncommon malformation seen in 0.1-0.25:10 000 of normal pregnancies [1]. The syndrome occurs more frequently in the offspring of diabetic than non-diabetic mothers [2]. The incidence is about one in 350 infants of diabetic mothers which is about 200-fold increase incidence in diabetic patient than general population [3]. Sacral [or lumbosacral agenesis in severe cases where lumbar spine is also involved] characterized by absence of the variable portion of the caudal portion of the spine. It is a very rare deformity. Patients with this deformity lack motor function at the affected vertebral level and sensory functions below the affected level.

Types
Renshaw classification divides the condition into four groups depending on amount of sacrum remaining and the characteristics of the articulation between the spine and the pelvis.

Type I: Partial or total unilateral sacral agenesis.
Type II: Partial sacral agenesis with a bilaterally symmetrical defect, a normal or hypoplastic sacral vertebra, and a stable articulation between the ilia and the first sacral vertebra.

Fig-1: Diagram shows partial unilateral sacral agenesis

Fig-2: Partial sacral agenesis
Type III: Variable lumbar and total sacral agenesis, with the ilia articulating with the sides of the lowest vertebra present.

Type IV: Variable lumbar and total sacral agenesis, with the caudal endplate of the lowest vertebra resting above either fused ilia or an iliac amphiarthrosis.

Fig-3: Variable lumbar and total sacral agenesis

Fig-4: Variable lumbar and total sacral agenesis

Type II: defects are most common, and type I are least common. Types I and II usually have a stable vertebral-pelvic articulation, whereas types III and IV produce instability and possibly a progressive kyphosis.

CASE REPORT
8 years old male patient came to the hospital unable to stand and move. His lower limbs are weak and there is a small degree of deformity. The clinical diagnosis is congenital hip dislocation.

X-ray pelvic was done and the diagnosis was complete absent of the sacral vertebrae (sacral agenesis) as shown in the radiograph below.

Fig-5: AP pelvic x-ray
DISCUSSION

With its speculative etiology, CRS remains associated with structural and systematic problems including genitourinary, gastrointestinal, orthopedic, neurological, respiratory and cardiac anomalies. Although there aren’t clearly determined causative factors, evidences have been found for maternal diabetes, genetic predisposition, and vascular hypoperfusion [4].

Some authors have supposed that CRS is a component of diabetic embryopathy. There have been many reports showing that maternal diabetes is associated with CRS [5, 6]. Several studies has reported partial genetic contribution of the development of the disease [2, 7]. A homebox gene, HLXB9 was supposed to be the major locus for dominantly inherited sacral agenesis [8]. But after a while, another study confirmed that the HLXB9 gene is not involved in the pathogenesis of CRS [9]. Since the normal spine requires close interaction between genes and environment, it is suggested that both genetic factors and teratogens may take part in pathogenesis of CRS. Some authors have supposed that there are critical stages of embryological development and several disorders take origin from the affected genetic structure by environmental factors during these periods [10].

Rojansky et al. have reported that pharmaceuticals, such as minoxidil, trimethoprim sulfamethoxazole, chemicals, fat solvents and appetite suppressants may play role in etiology of CRS [11]. Also, hyperglycemia, hypoxia, ketone or amino acid abnormalities have been suggested to be teratogens for infants of diabetic mothers [12].

REFERENCES


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