

Sacroccygeal Agenesis

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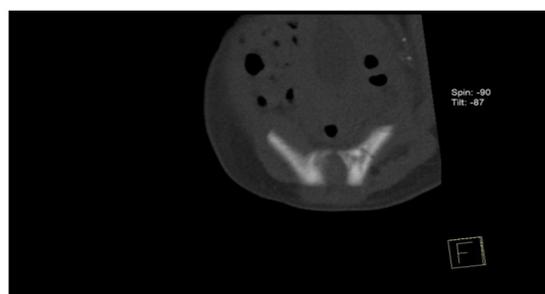
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CLINICAL HISTORY

A baby girl was born at 36 weeks gestation in Malala Maternity hospital with lower limb deformity (bilateral club-foot) and absent intergluteal cleft. Patient was referred for computed tomography of lumbosacral spine to rule out deformity of lumbosacral spine.

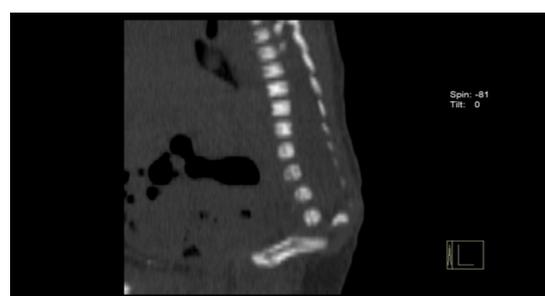
IMAGING FINDINGS

Computed tomography of lumbosacral spine demonstrated sacroccygeal agenesis. The two iliac bones are close to each other to replace the absent sacrum and coccyx. Patient also had bilateral club foot.



Area of Interest: Bones
Image origin: Radiology Department, FMIC
Sacroccygeal agenesis and closely situated two iliac bone

Figure 3: Axial CT bone window at the level of iliac bones shows sacroccygeal agenesis with closely situated two iliac bones.



Area of Interest: Bones
Image origin: Radiology Department, FMIC
sagittal image demonstrate sacroccygeal agenesis

Figure 4: CT bone window sagittal image demonstrate sacroccygeal agenesis.



Area of Interest: Bones
Image origin: Radiology Department, FMIC, Kabul, Afghanistan
Demonstrated sacroccygeal agenesis

Figure 1: VRT image demonstrate sacroccygeal agenesis.



Area of Interest: Bones
Image origin: Radiology Department, FMIC
Demonstrated sacroccygeal agenesis

Figure 2: VRT image demonstrate sacroccygeal agenesis.



Area of Interest: Bones
Image origin: Radiology Department, FMIC, Kabul, Afghanistan
Coronal image shows closely located iliac bones in the mid line.

Figure 5: CT Coronal bone window shows closely located iliac bones in the mid line.

DISCUSSION

Sacral agenesis was first described by Hohl in 1852 [1]. Sacral agenesis is rare entity occurring in 0.01% -0.005% of the population [2]. Duhamel coined the term caudal regression syndrome to describe a spectrum of congenital malformation; these range from simple anal atresia, to absence of sacral (sacral agenesis), lumbar and possibly lower thoracic vertebrae (caudal aplasia/dysplasia), to the most severe form with fusion of the lower extremities and major visceral malformation known as sirenomelia [3]. There is also an increased frequency of spinal cord abnormalities is seen in patients with sacral agenesis, including syrinx, tethered cord, lipoma, and lipomyelomeningocele [2].

Maternal Diabetes, genetic predisposition and vascular hypoperfusion have been suggested as possible causative factors [4]. A higher frequency, in the range of 0.1%-0.2%, has been observed in children of diabetic mothers [2]. Insulin injections may have a teratogenic effect on the embryo. This view has been supported by Duraiswami (1950) who has experimentally produced caudal regression in chickens by injecting insulin into the eggs [1].

Typically child present with flattened buttocks and shortened intergluteal cleft with absent lumbar lordosis. The pelvic ring is completed with either direct apposition of the iliac bones or with interposition of the lumbar spine replacing the absent sacrum. Scoliosis occasionally occurs. Visceral abnormalities such as anal atresia, fused kidneys and congenital heart malformations are not uncommon [1]. The spectrum of sacral agenesis can be categorized into four types.

In type 1, partial unilateral agenesis is localized to the sacrum or coccyx.

In type 2, there are partial but bilaterally symmetric defects in the sacrum. The iliac bones articulate with S1, and distal segments of the sacrum and coccyx fail to develop.

In type 3, there is total sacral agenesis and the iliac bones articulate with the lowest available segment of the lumbar spine.

In type 4, there is total sacral agenesis and the iliac bones are fused posteriorly along the midline [2].

The diagnosis is often made late in pregnancy. Early detection of caudal regression syndrome at 11 weeks of gestational age by transvaginal ultrasound scanning was reported. In the first trimester crown-rump length was found to be smaller than expected in caudal regression using abdominal ultrasound [4]. CT scan is not contributory enough and shows lumbosacral agenesis. MRI is the investigation of choice [5]. The management of these cases is symptomatic for each system, and the treatment should start as soon as possible [1].

FINAL DIAGNOSIS:

Sacrococcygeal Agenesis

DIFFERENTIAL DIAGNOSIS:

- Sacrococcygeal teratoma
- Meningocele

REFERENCES

1. Dounis E. (1978). Sacrococcygeal agenesis. A report of four new cases. *Acta Orthop Scand.* 49(5), 475-480.
2. Diel J, Ortiz O, Richard A, Losada, et al. (2001). The Sacrum: Pathologic Spectrum, Multimodality Imaging, and Subspecialty Approach. *Radiographics.* 21(1), 83-104.
3. Adra A, Cordero D, Mejides A, Yasin S, et al. (1994). Caudal regression syndrome: etiopathogenesis, prenatal diagnosis, and perinatal management. *Obstet Gynecol Survey.* 49(7), 508-516.
4. Aslan H, Yanik H, Celikaslan N, Yildirim G, et al. (2001). Prenatal diagnosis of Caudal Regression Syndrome: A case report. *BMC Pregnancy Childbirth.* 1(1), 8.
5. Jadav VJ, Gandhi J, Soni HC and Desai S. (2012). Caudal Regression Syndrome. *Gujarat Medical Journal.* 67(2), 129-131.