SACRAL AGENESIS

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INTRODUCTION:

Sacral agenesis (part of the caudal regression syndrome) is a rare and severe sacral developmental abnormality. It is a congenital malformation of unknown aetiology with possible involvement of genetic and teratogenic factors.Diagnosis is made clinically with prominence of the last vertebral segment and postural abnormalities. The associated malformations comprise anorectal, vertebral, urological, genital, and lower limb anomalies. Approximately 15-20% mothers of these children have insulin dependent diabetes mellitus.

REPORT:

A day 1 of life ,baby girl was referred to the orthopaedic department for abnormal lower limb posture.She was born via ELLSCS at 39w1d for 1 prev scar-refused VBAC,weighing 2.39kg, with an Apgar Score of 9/10.Her mother is a 42 y/o, para 3 with Overt DM on S/C Insulatard 8units ON and T. Metformin 1g BD. She was tolerating feeding well and was able to pass urine and motion.

Back examination showed a prominent lumbar vertebrae.Bilateral lower limbs were rigid with hips flexed up to 90 degrees, knees fully extended ,unable to extend the hips and flex the knees passively .The child also has bilateral club foot (PIRANI 6). Xray shows B/L ilium appearing to be fused. L4 and L5 vertebrae, sacrum and coccyx are absent.Ultrasound shows blunted and high Conus Medullaris at T10 level suggestive of tethered cord. No sonographic evidence of congenital renal anomaly.Subsequently, the baby was started on 4 hourly CIC. Serial Ponsetti casting was planned and she was referred to the occupational therapist and physiotherapist.

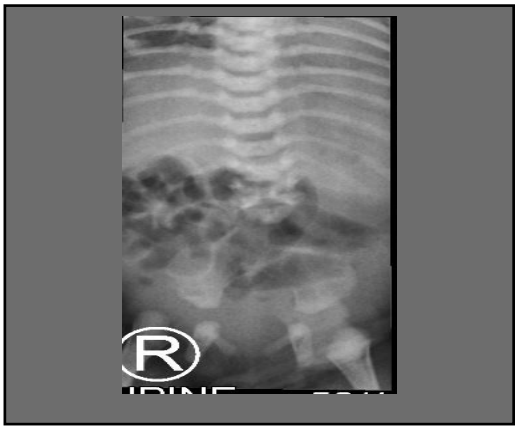

Figure 1: AP view of lumbosacral xray

Figure 2:Lateral view of lumbosacral xray

CONCLUSION:

The exploration and diagnosis of sacral agenesis is important in providing appropriate management. Management depends on the types of sacral agenesis (categorized by the affected spine and spinal cord morphology) and combined anomalies (mild life-long urological symptoms to severe gastrointestinal and musculoskeletal disabilities).

REFERENCES:1. Sacral Agenesis with Neurogenic Bladder Dysfunction—A Case Report and Review of the Literature ,Seema Sharma et al

2.MRI Findings of Type II Sacral Agenesis: A Case Report and Literature Review,Sang A Lee et al