Asphyxiating Thoracic Dystrophy

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Jeune first described asphyxiating thoracic dystrophy (ATD) in two siblings in 1954, which was subsequently delineated by several other reports. This skeletal dysplasia is characterized by contracted thorax leading to asphyxia neonatorum and repeated chest infections. We report a severe form of one such case.

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Case Report

A first male child of a non-consanguineous parents was delivered normally, developed respiratory distress within a few hours and was referred to M.S. Ramaiah medical teaching hospital. The mother was 26 years old with no antenatal risk factors. There was no family history of similar problems.

On examination, the baby was full term AFD. The baby's weight at birth was 2670 g, length 46 cm, head circumference 32 cm, arm span of 44 cm, with US:LS ratio of 1.9:1. The cry and activity of the baby was fair. He had signs of severe respiratory distress with bilateral crepitations. The cardiovascular system was normal. The liver was palpable three cm below the right costal margin. There was no facial dysmorphic features. The chest cage was long and narrow and the limbs appeared short.

Skeletal survey revealed classic features of ATD; the clavicles were highly placed; the thoracic cage was long and narrow with widening in the lower
region. Pelvis showed a trident acetabulum (Fig. 1). On ultrasound the kidneys were normal.

The baby was managed for respiratory distress with oxygen and IV fluids. Antibiotics were administered as sepsis screening was positive. The baby was subsequently seen for repeated respiratory infections and failure to gain weight.

**Discussion**

ATD is a rare autosomal recessive skeletal dysplasia with variable clinical, radiological and pathological manifestations. It is commonly subdivided into four clinical forms: lethal, severe, mild and latent based on the clinical course and radiological findings(1). The propositus belongs to the severe category. The classic infantile manifestations of ATD comprise of dwarfism with short limbs and characteristic radiographic changes in the ribs and pelvis(2). The reported case had all the classic clinical and radiological findings.

Though some of these infants present with respiratory distress and repeated chest infections, some would grow out of the disease and attain normal stature, though this would be difficult to predict during early infancy(3).

For those patients who get over the respiratory insufficiency during infancy, the renal involvement represents the main prognostic factor with the majority developing chronic renal insufficiency in the second decade of life(4). The histology shows features of nephronophthisis. Hepatic changes like portal tract fibrosis and bile duct proliferation have been described(3). The lungs are hypoplastic and some are dysplastic.

The management of these patients is primarily for respiratory distress and chest infections. Though some have resorted to reconstructive surgery of the rib cage the wisdom of such dramatic intervention is being questioned because of the high frequency of renal failure.

Prenatal diagnosis by ultrasonic measurement of the fetal length is possible which is useful in genetic engineering(5).
REFERENCES


Citrobacter Sepsis in Infants

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Gram negative bacilli are important agents causing neonatal sepsis(1,2). Infections may be acquired from the mother's genital tract or. nosocomial infections acquired from the delivery room or from the nurseries(3). Multidrug resistant Gram negative bacilli like Klebsiella, Citrobacter, Salmonella sp. and Pseudomonas are gaining importance in the recent years(1-4). Citrobacter species, recognized as saprophytes earlier are now known to cause neonatal meningitis, brain abscess, subacute bacterial endocarditis and sepsis(2,3). This communication presents clinical profile and antibiotic susceptibility pattern of five cases of citrobacter sepsis encountered in the pediatric wards of our hospital between the period June 1992 to May 1993.

Case Reports

Five cases of Citrobacter freundii were isolated from blood cultures of 2015 children (age group 0-1 year) with suspected sepsis, who were admitted in our hospital between June 1992 to May 1993. The total number of positive cultures were 480 (23.82%), out of which 5 were Citrobacter freundii (1.04%). Among the 5, two were neonates born in our hospital and 3 were outborn babies. Out of 5 cases, three were preterm neonates. The clinical signs and symptoms, complications, investigations and antibiotic sensitivity pattern of five infants suffering from citrobacter sepsis are given in Table I. All the 5 Citrobacter sp. were resistant to ampicilin, gentamicin,