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Sirenomelia (Mermaid syndrome) in an infant of a diabetic mother

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Abstract:

Caudal regression syndrome (Caudal dysplasia sequence) is a rare congenital malformation. It has a spectrum ranging from simple anal atresia to the absence of sacral, lumbar and possibly lower thoracic vertebrae and the most severe form called sirenomelia (Mermaid syndrome). Sirenomelia has a sole characteristic, which is the limbs fusion, with multiple internal structural abnormalities particularly in the renal tract (bilateral renal agenesis). This is a rare condition with a relative risk of 200-250 in diabetic pregnancies. The etiology of this syndrome is not well known. Maternal diabetes is considered to be other possible factors. We present birth of an infant with great congenital defect, which was categorized as the most intense form of caudal regression syndrome (sirenomelia). The baby was born from an uncontrolled diabetic mother who was ignorant of her diabetes. She had a sonographic report at early third-trimester of pregnancy, which had shown severe oligohydramnios and according to this reason the anomaly of the fetus was not detected at that time (antenatal). Since sirenomelia is a lethal abnormality, the infant died a few hours after birth. As notes above caudal regression syndrome is strongly associated with maternal diabetes; due to metabolic derangement in uncontrolled serum glucose.

Keywords:

Sirenomelia . Mermaid syndrome . Caudal regression syndrome . Caudal dysplasia sequence . Sacral agenesis

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