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Megacystis Microcolon Intestinal Hypoperistalsis Syndrome Case Report

KEYWORDS: Megacystis , Microcolon , Berdon , LSCS

Prof. S.V.Senthilnathan
Mch, Head of the Department, Department of pediatric surgery, Department of pediatric Surgery, Institute of child health and hospital for children, Egmore , Chennai

Dr.R. Ramesh Kumar
Resident , Department of pediatric Surgery, Department of pediatric Surgery, Institute of child health and hospital for children, Egmore , Chennai

ABSTRACT *: 3 days old ,girl child delivered by LSCS to a primi mother with Birth weight of 2.2 kg presented with history of abdominal distension and bilious vomiting, not passed of meconium. Antenatal Ultrasound findings shows that polyhydramnios, bilateral Ureterohydronephrosis with disdended bladder. On general examination cry and activity was good with generalized abdominal distension with palpable bladder. Child was investigated with imaging - Xrays (plain & Barium enema) and Ultrasound abdomen and diagnosed as a case of Megacystis microcolon intestinal hypoperistalsis syndrome. In view of poor prognosis , baby was managed conservatively with supportive measures only.*

Case History :
3 days old, Late preterm, girl baby delivered by caesarian section to a Primi mother with history of abdominal distension, bilious vomiting and not passed of meconium (fig .1). Antenatal ultrasound (fig.2)findings of polyhydramnios with bilateral ureterohydronephrosis with distended bladder. On examination child general conditioncry and activity was good with generalized abdominal distension and distended bladder.

Investigated with Imaging with plain X-ray abdomen (fig.3) shows paucity of gas shadow.

Barium enema findings of microcolon(fig.4). Ultrasound shows increased cortical echoes of Kidneys with distended bladder. With the above findings baby was diagnosed as a case of megacystis microcolon hypoperistalsis syndrome (Berdon syndrome).

Inview of the nature of the disease and poor prognosis, baby was managed conservatively with supportive care of parenteral nutrition and bladder drainage with catheter.

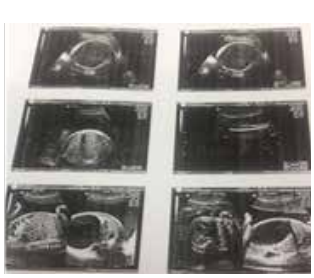
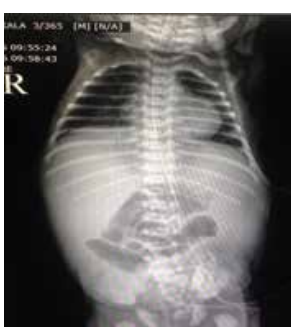


Figure 1. New born baby with abdominal distension With bladder catheter.
Fig 2. Antenatal USG Shows Distended bladder.



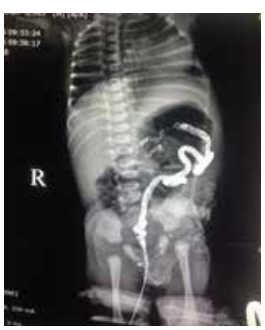


Fig 3. Plain x-ray abdomen shows Paucity of gas shadows.
Fig.4 Barium Enema with micro-colon.

Discussion :
Megacystis microcolon intestinal hypoperistalsis syndrome is a rare congenital and fatal disease. It is an autosomal recessive pattern with female predominant. The clinical features are abdominal distension with non-obstructive bladder , Micro-colon, hypo/ absent intestinal peristalsis. The associated malformations are incomplete intestinal rotation and short small bowel.Etiology for this syndrome is unclear . It could be either genetic /Neurogenic/Myogenic / hormonal . Pathogenesis are an imbalance in intestinal peptides , absence of interstitial

cell of cajal in the bowel and urinary bladder, vacuolar degenerative changes in the smooth muscle cells in the bowel and bladder. Genes involved in this syndrome are 15q24 & ACTG2.

Investigations :
Plain x- ray abdomen and Barium enema - paucity of gas shadow and microcolon are the characteristic features.

Ultrasonogram abdomen will demonstrate enlarged bladder with urterohydronephrosis. Intravenous urogram will show the bilateral uretero hydronephrosis with distended

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bladder.
Antenatal USG - Enlarged bladder & hydronephrosis, Polyhydramnios. From 16 week on wards - enlarged bladder Polyhydramnios from 3rd trimester onwards.

Amniotic fluid & Fetal urine analysis will shows increased digestive enzymes & calcium. Fetal MRI also useful investigation.

Management :
Management and the outcome of these babies are generally frustrating and poor. Surgery in the form of jejunostomy and cystostomy is generally needed. Nutritional Support is the mainstay of management. Eventhough the recent reports shows bowel transplantation has increased survival rate, these children needs lifelong immunosuppressive therapy and the prognosis is poor.

Conclusion : MMIHS is a very rare disease, it can be diagnosed in the antenatal Period itself , but the management of the disease is very difficult and majority of Children won't survive beyond one year of age. Recent trend shows intestinal transplantation may improve the survival rate , but in general the outcome is poor.

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