Megacystis Microcolon Intestinal Hyperperistalsis Syndrome (MMHS)

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Megacystis microcolon intestinal hypoperistalsis syndrome (MMIHS) is a rare and the most severe form of functional intestinal obstruction in the newborn. The major features of this congenital and usually lethal anomaly are abdominal distension, bile-stained vomiting, and absent or decreased bowel peristalsis. Abdominal distension is a consequence of the distended, unobstructed urinary bladder with or without upper urinary tract dilation. Most patients with MMIHS are not able to void spontaneously.
MMIHS

- Characterized by :-
  - Abdominal distension at birth,
  - Distended non-obstructive urinary bladder,
  - Bilateral hydronephrosis and hydroureters
  - Microcolon,
  - Short bowel with hypoperistalsis and malrotation, with hypoactive bowel sounds,
  - Ladd’s bands,
  - No ascites,
MMMHS

• Since Berdon et al. Reported 5 cases of MMIHS in 1976, there were only a 100 cases reported in the medical literature,
• A rare congenital disease,
• Found mainly in female infants,
• With high mortality rate,
• Etiology and pathogenesis are not clear,
• Most patients die within one year after birth from severe infection (recurrent UTI).
• Only few cases survived 4 to 11 years of age.
• Prenatal history of polyhydramnios,
• Physical examination at birth reveals a markedly distended abdomen with a large movable, ill-defined mass at the middle part of the lower abdomen (below the umbilicus),
• Other associate anomalies such as megaoesophagus, hydrometrocolpos and Barret’s oesophagus
Figure 1 Large fetal bladder seen on a longitudinal view of abdominal ultrasound at 22 weeks gestation. The fetus is in prone position.
MMIHS

- Investigations:
  - Chest, abdomen, bone survey x-rays, (megacystis, hydronephrois, hydroureters, high intestinal obstruction possibly only distended stomach),
  - BUN and Serum Creatinine elevated, low serum calcium
  - BG, metabolic acidosis,
Figure 3 A contrast enema showing microcolon in an MMIHS patient.
MMIHS

• **Investigations :-**

  • Voiding cystourethrography, shows markedly distended bladder, smooth walled without intrinsic or extrinsic lesions,

  • Upper GI study shows distended stomach with normal gastric contour with or without doudenal cap,
Figure 2 Voiding cystourethrogram showing a massively enlarged bladder in an MMIHS patient
MMIHS

- **Surgical procedures required,**
  - Laparotomy, malrotation, microcolon, Ladd’s bands, short intestines with hypoperistalsis, huge urinary bladder. Required Ladd’s procedure, ileostomy, and cutaneous vesicostomy
  - Bowel biopsy ileal, colon, sigmoid,
  - Rectal biopsy
  - Central line.
Figure 4
Operative photograph of a massively dilated urinary bladder in MMIHS.
Radiologic evaluation usually suggested the diagnosis of MMIHS. Plain abdominal films showed either dilated small bowel loops or a gasless abdomen with evident gastric bubble. An enlarged urinary bladder was present in all patients who had cystography or ultrasonography. Cystography showed vesicoureteral reflux Intravenous urography or ultrasonography detected unilateral or bilateral hydronephrosis.
MMIHS

- The most important prenatal ultrasonographic finding is distended urinary bladder without oligohydramnios which is different from obstructive uropathy disease,
- The earliest time to detect distended urinary bladder and hydronephrosis is 16-20th week of gestation,
- Also distended stomach, increased amniotic fluid, dialted renal calyces and megaureter can can be viewed.
MMIHS

• **Differential diagnosis :-**
  • Depends on the main manifestations,
  • For megacystis ; from prune belly syndrome, urethral agenesis, and variant of caudal regression are needed,
  • For microcolon; from intestinal atresia, volvulus, and total colon aganglionosis are needed,
  • For ileus or pseudoobstruction; from hypothyroidism, hypokalemia and maternal narcotic drugs are excluded.
MMIHS

- **Treatment :-**
  - Nutritional support is the main stay of treatment. Hence hyperalimentation is required,
  - Paliative surgery such as jejunostomy, ileostomy or cystostomy are required,
  - Prokinetic drugs for promoting bowel motility are ineffective,
  - ?? Multiviscerel transplantation,
Outcome :-
The management of patients with MMIHS is frustrating. A number of prokinetic drugs and gastrointestinal hormones have been tried without success. Surgical manipulation of the gastrointestinal tract has generally been unsuccessful. The need for surgical intervention should be made carefully and individualized, in that most explorations have not been helpful and probably are not necessary.
Prognosis :-
The overall prognosis is considered to be poor and
treatment has been shown to be generally
ineffective $^6$.
It is usually lethal within the first year of life $^3$.
Megacystis and microcolon were the two most
frequent findings at surgery or autopsy
MMIHS

Associations:
- polyhydramnios: can be present in ~25% of cases
- omphalocoele
- intestinal malrotation \(^{10}\)