

What is MMIHS?

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Megacystis Microcolon Intestinal Hypoperistalsis Syndrome (MMIHS, Berdon Syndrome) was first described by Walter Berdon in 1976. It is an extremely rare disorder that affects the bladder and gastrointestinal tract. Kidney complications (secondary to bladder complications) are also common.



Prior to diagnosis or treatment, most patients with MMIHS present with all or some of the following indicators:

- a large, unobstructed bladder
- difficulty or inability to void urine
- a small colon
- intestinal malrotation
- decreased intestinal motility and nutrient absorption



Education and awareness of MMIHS is critical as proper diagnosis and treatment can significantly impact the prognosis for each individual.

Please visit our website to learn more and join the effort!

For more information

www.mmihs.org

please visit:

The creation of this brochure is dedicated to all those who have been touched by this rare syndrome. May our efforts contribute to making the future a place that is as extraordinary as each of you.



MMIHS



MEGACYSTIS

an unusually enlarged bladder

MICROCOLON

a small colon



INTESTINAL HYPOPERISTALSIS

abnormally sluggish intestinal muscular contractions



SYNDROME

a condition characterized by a set of associated symptoms



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Diagnosis

MMIHS is typically diagnosed prenatally or shortly after birth. The gastrointestinal complications of MMIHS can sometimes be mistaken for Hirschsprung's disease. However analysis of a colonic biopsy will show the presence of ganglion cells (which are not present in Hirschsprung's patients), and in some cases there may be an abundance of these cells.

Prenatal Diagnosis

The leading indication of MMIHS in utero are:

- an enlarged fetal bladder (megacystis)
- bilaterally enlarged kidneys (hydronephrosis)
- undescended testicles (in male patients)
- Polyhydramnios, or increased amniotic fluid in late pregnancy
- decreased or absent fetal colon activity in the third trimester.

Postnatal Diagnosis

Post birth indicators may include:

- a distended abdomen
- enlarged bladder and kidneys as evident on neonatal ultrasound
- bile stained vomiting
- failure to pass meconium

A diagnosis of MMIHS can be confirmed with genetic testing.

Symptom Management



Diligent care on the part of medical professionals and caregivers is critical. Treatment involves targeting the bladder, kidney and intestinal complications.

Bladder and Kidney Treatment

Most patients with MMIHS have a bladder that is unable to empty on its own either at all, or as regularly as it should. For this reason, it is critical that a bladder-emptying strategy be in place to keep bladder pressure low and prevent reflux into the upper urinary tract.

Catheterization along with possible use of antibiotics which decrease the risk of frequent Urinary Tract Infections (UTI) seems to be the optimal and most commonly used strategy.

Intestinal Treatment

Intestinal malrotation and lesions can be common in MMIHS patients and can be corrected with surgical intervention. However, the complications related to Intestinal Hypoperistalsis is considered the more serious and difficult symptom to manage.

The primary goal of treatment is to provide adequate nutrition and limit abdominal distention caused by slow intestinal motility. Patients with MMIHS often have surgery to create an ileostomy and/or intestinal resection to help minimize abdominal bloating and improve intestinal motility. A feeding tube is also commonly placed to maximize fluid and nutrient intake.

Unfortunately, for most MMIHS patients, surgical interventions alone are not able to allow their bodies to digest enough food to get the nutrients that they need. For that reason, to some degree, a majority of patients with MMIHS rely on Total Parenteral Nutrition (TPN). A central line, along with the feeding tube are some of the "accessories" that help MMIHS patients lead a more "normal" life.