

# Obstructed umbilical hernia in a child with Hurler's syndrome

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## ABSTRACT

Hurler's syndrome (MPS I-H) is a lysosomal storage disorder characterized by physical deformities and developmental anomalies and premature death. Deficiency of the enzyme alfa-L-Idronidase results in intralysosomal accumulation of dermatan sulfate and heparan sulfate resulting in cell dysfunction. It manifests as progressive hepatosplenomegaly, cardiac disease, severe skeletal abnormalities, hydrocephalus and mental retardation. Umbilical and inguinal hernias are common in this disorder. Obstruction of hernia in these children is rare because of wide hernial ring. This is a discussion on a child with Hurler's syndrome who presented with obstructed umbilical hernia. Surgery was performed under general anaesthesia after relieving the obstruction conservatively. Surgical and anaesthetic problems in this syndrome are discussed in the present communication.

**Key Words:** Hurler's syndrome, Mucopolysaccharidoses, Umbilical hernia, Mayo's repair

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## INTRODUCTION

Hurler's syndrome is a rare lysosomal storage disorder in which mucopolysaccharides, dermatan sulfate and heparan sulfate are accumulated in the lysosomes causing cellular dysfunction. The incidence is 1:1,00,000 births.<sup>[1]</sup> Patients with Hurler's syndrome have many medical problems, including progressive developmental delay, airway obstruction, cardiac disease, hepatosplenomegaly, and severe joint restriction, and most die by the age of 10 years.<sup>[1]</sup> Umbilical or inguinal hernias are constant features but obstruction of umbilical hernia is rare.<sup>[2]</sup> Anaesthesia and surgery in these children is a challenge. We report our experience of the surgical and anaesthetic management of obstructed umbilical hernia in a patient with Hurler's syndrome and discuss the various anaesthetic and surgical problems faced in this syndrome.

## CASE REPORT

A 15-year-old girl, a known case of Hurler's syndrome, presented with umbilical hernia since one year of age. Hernia became irreducible along with symptoms of obstruction 3 months before the present admission. On examination the child was short. Her height was 98 cm and weight 16 kg, which were less than the 5<sup>th</sup> percentile on the NCHS standard. Features of Hurler's syndrome like large head, coarse facial features, depressed nasal bridge, large thick tongue, hazy cornea, short neck, joint contractures, kyphosis, wide ribs and pectus excavatum (Figure 1), were obvious on clinical examination. Radiological examination showed features of dysostosis multiplex that included large skull with thickened calvarium, enlarged J-shaped sella, abnormal spacing of teeth, anterior hypoplasia of lumbar vertebrae, oar-shaped ribs, poorly formed pelvis with small femoral heads and coxavalga. The child had umbilical hernia around 10 cm in diameter with a narrow base 3 cm in diameter, and it was irreducible. The skin over the hernia showed ischaemic changes with hyperpigmentation (Figure 2). Cardiac auscultation showed Grade III systolic murmur. Her IQ was 50 indicating moderate mental retardation.

Hematological investigations were within normal limits. Echocardiogram showed thickened mitral valve and

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**Figure 1:** Child with Hurler's syndrome with obstructed umbilical hernia—preoperative photograph.



**Figure 2:** Obstructed umbilical hernia with ischaemic skin changes

interventricular septum. LV function was normal. The child was treated conservatively and the obstruction was relieved in two days. Surgery was done under GA for the hernia, in a planned manner, at the earliest. Small bowel of about one foot was found as the content of the hernial sac. A part of the bowel was ischaemic but viable. A defect of 3x3 cm was repaired by Mayo's technique. The postoperative period was uneventful.

## DISCUSSION

Mucopolysaccharidoses are a heterogeneous group of inheritable storage diseases characterized by intralysosomal accumulation of glycosaminoglycans (GAGs), excessive urinary excretion of GAGs and progressive mental and physical deterioration. Hurler's

syndrome (MPS I-H) is the prototype of MPS.<sup>[3]</sup> Hurler's syndrome is a lysosomal storage disease caused by a deficiency of alfa-L-iduronidase, resulting in interference with cellular function because of excess accumulation of partially degraded GAGs like dermatan sulfate and heparan sulfate within the cells.<sup>[1]</sup>

At diagnosis, which is usually is made between 6-24 months of life, clinical features usually noted are hepatosplenomegaly, skeletal deformity, coarse facial features, corneal clouding, large tongue, prominent forehead, joint stiffness and short stature. Other constant features are inguinal and umbilical hernias, large and scaphoid head, small widely spaced teeth, noisy respiration, and limited joint mobility. Later signs include cardiac murmurs, deafness, blindness and short stature. The constellation of skeletal abnormalities is known as dysostosis multiplex.<sup>[2]</sup> When the MPS disorder is suspected based on clinical features, radiographic results or urinary screening tests, definitive diagnosis is established by enzyme assay.<sup>[3]</sup>

We faced problems related to anaesthesia and postoperative care. Hurler syndrome is the worst airway problem in paediatric anaesthesia.<sup>[4]</sup> Odontoid dysplasia and radiographic subluxation of C1 on C2 is common. This may cause anterior dislocation of the atlas and spinal cord compression. Precaution should be taken to prevent unnecessary flexion or extension of the neck.<sup>[4]</sup> Spinal anaesthesia is contraindicated because of spinal anomalies. Thickening of the soft tissues, enlarged tongue, short immobile neck and limited mobility of the cervical spine and temporomandibular joints make laryngoscopy and intubation difficult.<sup>[5]</sup> Spinal deformities, hepatosplenomegaly and recurrent lung infections may inhibit pulmonary function. Nasal airway is more effective but advancement is difficult because of mucopolysaccharide deposits.<sup>[5]</sup> Fibreoptic laryngoscopy and bronchoscopy may be used for intubation. In difficult cases the laryngeal mask airway has proved to be a useful additional aid.<sup>[6]</sup> In severely affected patients, the clinical course is dominated by frequent upper and lower respiratory infections.

Although seen in healthy infants, a number of clinical disorders are associated with umbilical hernias, including trisomy 21, congenital hypothyroidism and mucopolysaccharidosis. Most hernias close by 3-4 years.<sup>[7]</sup> Hernias with diameter >2 cm are less likely to close.<sup>[7]</sup> Unlike inguinal hernia, incarceration or strangulation in umbilical hernia is rare during early childhood. Obstruction of hernia is rare in mucopolysaccharidoses because of the wide ring. Hernias are repaired surgically for the prevention of obstruction or strangulation. Traditionally, Mayo's overlap repair is used. Large hernias can be repaired by an onlay nylon darn of anterior rectus sheath,

prosthetic mesh repair or shoelace repair.<sup>[8]</sup> Common surgical procedures performed in these children include indwelling CVP placement, herniorrhaphy, VP shunt placement, ENT procedures, spine surgeries, carpal tunnel repair, dental restorations and cholecystectomy.<sup>[4]</sup>

Surgeons operating for various indications in children with developmental deformities should be aware of common surgical and anaesthetic problems and safe surgical procedures can be performed with due care being taken.

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