

Giant Umbilical Hernia in a Patient with Hurler's Syndrome: Case Report and Management Principles

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Abstract

The surgical and anaesthetics management of patients with giant umbilical hernia associated with Hurler's Syndrome is a challenge because of many medical problems including progressive developmental delay, the observation that we report proves that the umbilical hernia that accompanies the Hurler's syndrome may reach impressive size with a risk of complication; Spinal anaesthesia is contraindicated because of spinal anomalies.

Laryngoscopy and fibreoptic bronchoscopy may be used for intubation in difficult cases the laryngeal mask airway has proved to be a useful additional aid

Despite the risk, it may be wise to perform elective hernia repair using a prosthetic mesh in selected cases of Hurler's syndrome.

Keywords: Hurler's syndrome; Giant Umbilical Hernia; Mucopolysaccharidosis

Introduction

Hurler's syndrome is a genetic disorder that results in the buildup of glycosaminoglycans (formerly known as mucopolysaccharides) due to a deficiency of alpha-L iduronidase

The surgical and anaesthetics management of patients with giant umbilical hernia associated with Hurler's Syndrome is a challenge because of many medical problems including progressive developmental delay, dysmorphic facies, airway obstruction, cardiac disease, hepato-splenomegaly and severe joint restriction [1]. Fiberoptic laryngoscopy will be used for intubation because the abnormal laryngeal anatomy.

Umbilical and inguinal hernias are common in this disorder and generally treated conservatively, because complications such as incarceration are rare, and risks involved in surgical correction are high [2].

We report our experience and discuss the various anaesthetic and surgical problems in the management of a giant umbilical hernia in a patient with Hurler's syndrome.

Case Report

14 years-old girl with Hurler's syndrome who presented with obstructed giant umbilical hernia around 18 cm in diameter with ischemic skin changes (**Figure 1**).

Abdominal CT-Scan and X-Ray were realized: showed a hug umbilical hernia with fascia defect of 08 cm (**Figure 2**).



Figure 1 Giant umbilical hernia in a patient with Hurler's syndrome After relieving the obstruction conservatively, hernia was repaired under general anaesthesia. Regular intubation failed as a result of abnormal laryngeal anatomy, necessitating fiberoptic laryngoscopic intubation.

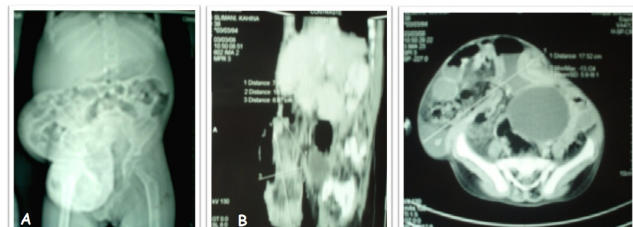


Figure 2 A: Abdominal X-Ray / B: Abdominal Scan

Through a transverse skin incision, the hernial sac was dissected free. The defect of the fascia (8cm) was closed primarily with overlapping layers using non absorbable sutures with insertion of a prosthetic mesh.

The postoperative course was uneventful, and the patient was discharged 15 days after admission. Six months after the operation the patient was doing well with no evidence of recurrence (**Figure 3**).



Figure 3: Postoperative photograph

Discussion

The observation that we report proves that the umbilical hernia that accompanies the Hurler's syndrome may reach impressive size with a risk of complication, which encourages us to reconsider the attitude abstentionist in some cases.

The arguments that have prompted a surgical treatment are mainly represented by the volume of the hernia, the real risk of strangulation and the important functional impairment [3].

Operative treatment may result in a laborious course. Airway management can be difficult because of the large

tongue and the short neck with limited movement [4]. The trachea may be narrow, the soft tissues are swollen and bleed easily, and these patients are sensitive to bronchospasm.

Spinal anaesthesia is contraindicated because of spinal anomalies.

Laryngoscopy and fibreoptic bronchoscopy may be used for intubation. In difficult cases the laryngeal mask airway has proved to be a useful additional aid [5].

We chose to use a prosthetic material in primary hernial repair to avoid the risk of recurrence. A mesh graft was safely used to support the patient's abdominal wall.

Ethical approval

Necessary approval was taken from the Institution and the patients for carrying out this work.

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