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Congenital Duodenal Diaphragm in association with Situs Inversus: A Case Report

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Summary

Nzeh DA. Congenital Duodenal Diaphragm in association with Situs Inversus: A Case Report. *Nigerian Journal of Paediatrics* **1986; 13:91.** The occurrence of congenital duodenal diaphragm in association with situs inversus in a neonate who also had glucose-6- phosphate dehydrogenase deficiency is reported. The need to look for possible associated anomalies in cases of congenital duodenal diaphragm, is emphasized.

Introduction

THE duodenal diaphragm or web is a rare congenital anomaly.[1] Diaphragms occur most commonly in the second portion of the duodenum and can be an unusual cause of intestinal obstruction in neonates and adults alike.[2] Congenital anomalies that are known to be associated with duodenal diaphragms include congenital heart disease, Down's syndrome, malrotation of the gut and situs inversus.[3]

The purpose of the present communication is to draw attention to the likelihood of associated congenital anomalies in cases of congenital duodenal diaphragm as well as highlight the usefulness of barium meal in localizing the site and nature of the obstruction, prior to surgery.

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Case Report

BO, a one-month old female child, was admitted to hospital with a history of persistent projectile vomiting since the age of four days. She was ill-looking, wasted and had abdominal distension. Barium meal examination revealed a right-sided stomach and an obstruction to the transit of barium between the first and second parts of the duodenum. The stomach and the outlined portion of the duodenum showed marked distension. A rounded configuration was noted at the site of obstruction in the duodenum (Fig 1). Two-hour delayed radiograph of the abdomen after barium meal, showed minimal emptying of barium from the stomach (Fig 2). The baby was also discovered to have glucose-6-phosphate dehydrogenase deficiency.

Laparotomy was performed to relieve the duodenal obstruction; situs inversus was confirmed at laparotomy. The bulk of the liver was located to the left of the midline while the

Fig 1. Barium meal showing a distended and right- sided stomach. The outlined duodenum has a rounded configuration at the site of obstruction.

stomach and spleen were lying to the right of the midline. The spleen was multilobular while the stomach was distended and hypertrophied. A duodenal diaphragm was found in the first part of the duodenum and was excised. Unfortunately, the child died five days after surgery.

Discussion

Although the precise aetiology of congenital duodenal diaphragm is unknown, it is believed that vascular accidents *in-utero*, particularly those that interfere with mesenteric blood supply may be responsible[4]. The incidence of congenital duodenal diaphragm has been put at 1 in every 9,000 to 40,000 births[1]. Development of clinical symptoms appear to be inversely related to the size of the diaphragmatic aperture. Vomiting is a consistent symptom. Depending on the location of the web in relation to the ampulla of Vater, vomitus may or may not be bile stained.

The plain abdominal radiograph may be unremarkable but a varying degree of obstruction at the duodenal level may show a classical "double-bubble" appearance due to gaseous distension of the stomach and duodenum. Although this sign is not diagnostic, it is taken as the most common radiological finding in transverse duodenal diaphragm especially in the neonate[5]. Barium meal shows the site of obstruction in the duodenum in most cases and may sometimes, appear rounded, due to prolapse of the diaphragm, as occurred in the present case. Duodenal dilatation and prolapse of the web are accentuated in neonates who had severe obstruction in foetal life as in the present case or in older children[5].

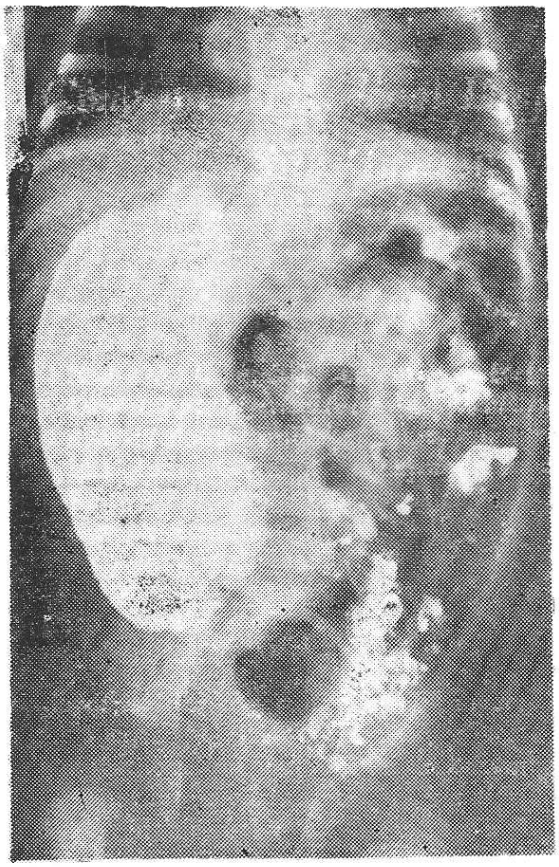

Fig 2. Two-hour delayed film following barium meal. The stomach shows minimal emptying of barium. The apex of the heart is seen pointing in an opposite direction to the stomach.

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