Modification of the endoscopic management of congenital duodenal stenosis

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This report documents a new endoscopic management modality for congenital membranous stenosis in the third portion of the duodenum. Standard approaches to duodenal stenosis in newborns include a laparotomy with an enteroenterostomy, bypassing the obstruction, or a duodenoduodenostomy with excision. We successfully developed a modification of the endoscopic treatment modality for congenital duodenal diaphragm.

Key words: congenital duodenal stenosis, endoscopy.

Congenital duodenal diaphragm is a rare condition of intestinal obstruction, with an incidence of approximately 1 in 10,000[1]. In a large series, the ratio of atresia:stenosis was also 3:2 or 2:2, although the incidence has been variously estimated as ranging from 1 in 10,000 to 1 in 40,000[2]. The pathogenesis of such a malformation is an incomplete resorption of the duodenal epithelial ingrowth during embryonic life[3]. Duodenal diaphragms or webs with or without central openings in neonatal patients can usually be dealt with by either a duodenoduodenostomy or excision[2]. This is a case report of a nine-day-old infant with duodenal diaphragm who was successfully treated with a modification of the endoscopic treatment modality.

Case Report

A 31-year-old woman, gravida 3, para 1, abortion 2, was referred to the Maternity and Perinatal Care Center of Fukuoka University Hospital at 18 weeks' gestation because of polyhydramnios and a distended stomach and duodenum of the fetus, thus suggesting congenital duodenal atresia. She had no particular medical complications. The fetus demonstrated no other anatomical defects. The patient underwent amniocentesis, which demonstrated the normal 46, XY chromosomes. Her husband had a past history of congenital duodenal atresia. Although the distended stomach and duodenum were continuously present following the fetal ultrasonographic examination (Fig. 1), polyhydramnios disappeared at 35

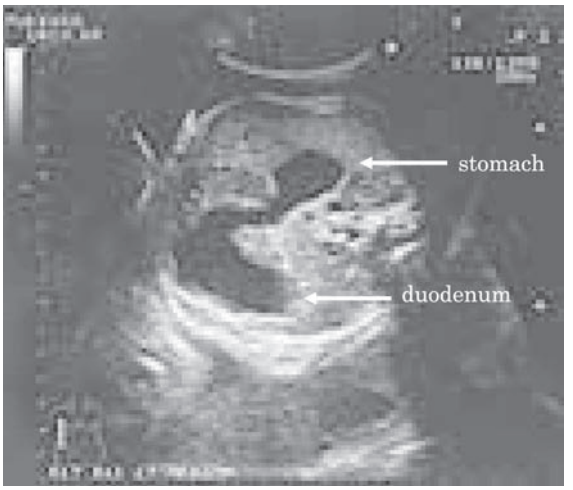

Fig. 1. An ultrasound scan of the fetus revealed a distended stomach and duodenum.

weeks' gestation. These findings thus suggested the fetus to have congenital duodenal stenosis either with or without a diaphragm.

A male of 39 weeks' gestation weighing 3,327 g was born by a spontaneous vaginal delivery. The Apgar score was 9 at both 1 and 5 minutes. An abdominal X-ray demonstrated a dilated stomach, an abnormal course of air-filled duodenum to the right and a very small amount of air in the intestinal tract. An upper gastrointestinal series revealed duodenal stenosis to be present with a dilated third portion of the duodenum and a pin hole. The boy underwent an endoscopic examination with an Olympus GIF-N30 (5.2 mm in diameter: Olympus, Tokyo, Japan) and re-upper gastrointestinal series under general anesthesia at nine days of age, when his body weight was 3,060 g (Fig. 2). The mucosal diaphragm in the third portion of the duodenum showed an opening like a pin hole with mucous bubble under endoscopic observations (Fig. 3a and b). The papilla of Vater was identified on the oral side of the stenosis. First, the surrounding pin hole of the diaphragm was dilated by an ablation of Argon Plasma Coagulation (APC) (ABC300; Erbe. Tuebingen, Germany). After a guide wire was placed though the dilated pin hole (Fig. 3c), the hole was then dilated with a 12-mm balloon catheter. After removing the dilator, the endoscope was then reinserted and a lumen measuring approximately 8 mm in diameter was demonstrated through the diaphragm (Fig. 3d). No complications were observed.

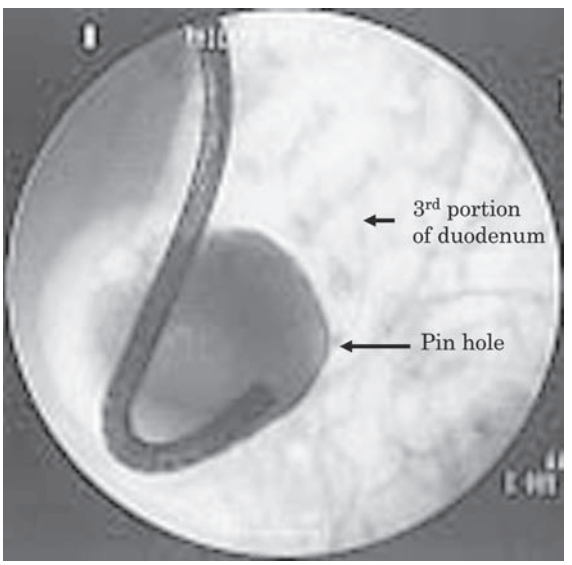

Fig. 2. Upper gastrointestinal series shows a dilated second portion of the duodenum with stenosis resembling a pin hole.

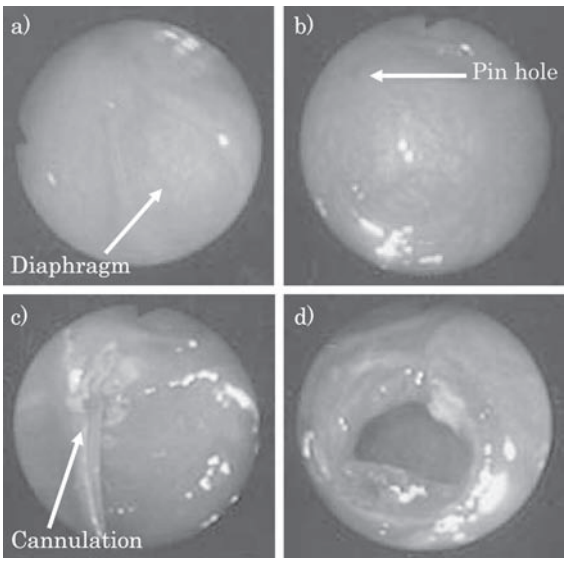

Fig. 3. Endoscopic view of the diaphragm in the third portion of the duodenum: (a) mucous bubble (diaphragm), (b) very small opening like a pin hole, (c) a JAG wire placed though the dilated pin hole, and (d) the lumen measuring approximately 8 mm in diameter after the removal of the dilator.

At 30 and 51 days of age, balloon dilatations were performed twice more using 13 and 13.5-mm balloon catheter due to the infant's continued inability to tolerate oral feeding, and a contrast study of the upper gastrointestinal tract showed a new reconstruction of the lumen. After dilation, the patient finally began to gradually tolerate a liquid diet. Seven months later, repeat endoscopy demonstrated no stricture of the duodenum. He could begin to tolerate a soft diet. He is now 16 months old and doing well.

Discussion

Congenital duodenal strictures rank third after an imperforate anus and esophageal atresia among cases of intrinsic obstruction of the alimentary tract in newborns[4]. Despite this, duodenal webs or diaphragms are rare[1,2]. However, the etiology is unknown[3,5,6] and there are no known associations[5]. Duodenal diaphragm consists of the mucosa and submucosa, and it rarely has a muscular layer[5,6]. Changes in the duodenal motility with advancing age, along with a

disproportion between the increase in size of the duodenal lumen and size of the aperture are thought to be important[6,7].

The diagnosis of duodenal diaphragm in infants and children is often difficult. Traditional upper gastrointestinal study is notoriously insensitive and they are often repeatedly negative. Its sensitivity may increase when hypotonic duodenography is also used. When positive, it shows proximal duodenal dilatation and distal obstruction[4], similar to the findings shown in Fig. 2. Endoscopically, the lesion appears as an annular web with an often acentric opening ranging from 2 to 8 mm in diameter[4]. The narrow, nipple-shaped lumen with a circumferential cuffing of duodenal mucosa has been compared with the female anatomy and dubbed the "duodenal cervix sign" by Halko et al.[8]. About one and a half years ago, we experienced another case of duodenal membranous stenosis in a Down syndrome infant. The patient underwent a laparotomy and endoscopic observation to determine the true state of the diaphragm and a small opening was found. The diaphragm was a wavy mucous bubble. In the present case, it was very difficult for us to identify a pin hole in the diaphragm. Therefore, we performed duodenography and endoscopic observations simultaneously. As a result, we were able to identify the hole owing to our previous experience.

The standard operative approach for a correction of congenital duodenal obstruction caused by stenosis is a laparotomy with an intestinal bypass or duodenotomy with an incision or excision of the obstruction[1,9]. The development of new technology now allows us the option of a safe, effective and new treatment[9]; for instance, endoscopic equipment and instruments have all markedly improved[10]. A dilatation of duodenal strictures in adults is usually attempted using a dilatation balloon[1,11]. Some other authors have reported the removal of a diaphragm using electrocautery papillotome or an endoscopic laser in adults[1,3,4,6,9,11].

Because of the limitations imposed by the size of the fiberscope, it is very difficult to perform endoscopic treatment in infants. Okamatsu et al.[10] reported that a 60-day-old baby boy with congenital membranous stenosis was successfully treated with an endoscopic membranectomy by means of a high-frequency wave cutter, following balloon dilatation. The Nd: YAG laser[12] and KTP/532 laser[9] have also been used for endoscopic laser ablation of duodenal webs in children. However, the Nd: YAG laser did not effectively perforate the duodenal web of our previous congenital duodenal membranous atresia case associated with Down syndrome. Additionally, tangential irradiation is not possible with this method, and this is one obvious drawback[13]. The Nd: YAG laser has a depth of tissue penetration of 1-4 mm, such that the coagulative necrosis resulting from the application of these devices may result in an undesirable intestinal perforation or damage to structures outside the lumen[9].

In recent years, Grund et al.[14] utilized APC to reduce the tumor mass, treat tumor ingrowth within a stent, and release tumor compression. APC is a thermal coagulation technique that uses ionized argon to transmit high-frequency electrical current, contact- free, to tissue, making it possible to provide tangential irradiation to coagulate a target site uniformly. This feature is one advantage over conventional endoscopic cauterization[13]. Sagawa et al.[13] revealed that the depth of coagulation using APC could easily be adjusted by changing the irradiation time and current output in an ex vivo experiment using gastric mucosal tissue, thus suggesting that this technique is both safe and effective for human application. Therefore, we used the ablation of APC for the dilatation of the surrounding pin hole of the diaphragm, and thereafter we performed balloon dilation.

No report has been published to date describing the use of APC as an effective treatment of duodenal diaphragm. Although a few infantile cases of endoscopic management of duodenal strictures have been reported[9,10,12], including our case, we believe that this procedure is an excellent alternative to surgical management. However, more experience is needed to further delineate the exact morbidity and mortality associated with this treatment.

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