

## Case Report

### Laparoscopic Repair of Congenital Duodenal Obstruction

CONSTANTINE T. FRANTZIDES, MD, PhD,<sup>1</sup> ATUL K. MADAN, MD,<sup>2</sup>  
PUNJ K. GUPTA, BS,<sup>3</sup> CLAIRE SMITH, MD, FACR,<sup>4</sup> and ALI KESHAVARZIAN, MD<sup>5</sup>

#### ABSTRACT

**CHARGE syndrome (or association) refers to a group of physical abnormalities occurring together: coloboma, heart defect, atresia choanae, retarded growth and development, genital hypoplasia, and ear anomalies/deafness. We report the successful use of laparoscopy in a patient with CHARGE syndrome and congenital duodenal obstruction.**

#### INTRODUCTION

LAPAROSCOPIC TREATMENT OF MULTIPLE DISEASE PROCESSES continues to evolve. Avoiding a large incision in the abdomen and its associated complications are benefits of laparoscopy in many operative situations. One such situation is intestinal obstruction, whether from adhesions from previous surgery or from congenital abnormalities. Laparoscopy has been previously used in the treatment for malrotation in adults.<sup>1</sup> We report the successful use of laparoscopy in a patient with CHARGE syndrome and congenital duodenal obstruction.

#### CASE REPORT

A 23-year-old female with CHARGE syndrome had a history of intermittent nausea and vomiting since birth. The patient had nonbilious emesis with no other major complaints. An upper gastrointestinal barium study showed a redundant, tortuous, and partially obstructed postbulbar duodenum (Fig. 1).

The patient was offered a diagnostic laparoscopy. Four ports were placed: infraumbilically, at the right lower quadrant, at the left lower quadrant, and at the right upper quadrant. The duodenum had a redundant intraperitoneal first portion with multiple bands attached that were causing obvious kinking of the duodenum. These bands were carefully dissected. A Kocher maneuver was performed to assure that all the bands had been taken down. No kinking of the duodenum noted and the patient was extubated and transferred to the recovery room with no complications.

On postoperative day 2, the patient had an upper gastrograffin swallow, which showed the duodenum in the normal position, with a redundant hammock-shaped deformity. There was minimal thickening of the folds in this portion of the duodenum, with the duodenal lumen patent, with free passage of contrast material through the duodenum into the proximal jejunum (Fig. 2).

By postoperative day 3, the patient was comfortable, tolerating her feeds, had bowel movements, and ambulating; she was discharged that evening. On one-year follow-up, she was free of nausea or vomiting symptoms.

F1

F2

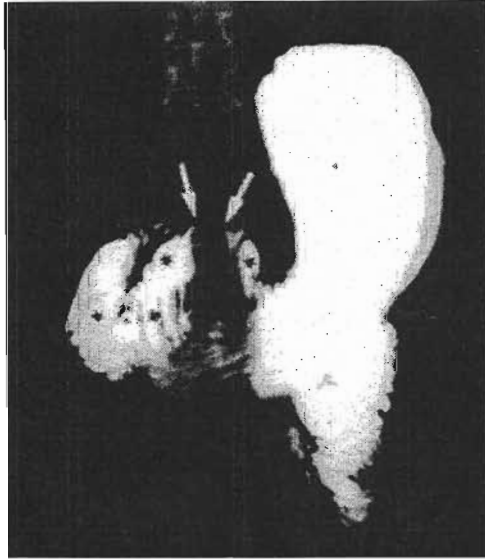
<sup>1</sup>Department of Surgery, Evanston Northwestern, Evanston, Illinois.

<sup>2</sup>Department of Surgery, University of Tennessee Health Science Center, Memphis, Tennessee.

<sup>3</sup>Department of Surgery, Rush University, Chicago, Illinois.

<sup>4</sup>Department of Diagnostic Radiology and Nuclear Medicine, Rush University Medical Center, Chicago, Illinois.

<sup>5</sup>Section of Gastroenterology and Nutrition, Department of Medicine, Rush University, Chicago, Illinois.



**FIG. 1.** The preoperative contrast exam shows the duodenum (asterisks) to be redundant and a focal narrowed contour (long white arrows). The ligament of Treitz is obscured by the stomach.

## DISCUSSION

CHARGE syndrome is a collection of congenital conditions first described by Hall,<sup>2</sup> occurring at an estimated rate of 1 in 10,000 infants.<sup>3</sup> CHARGE refers to the physical abnormalities occurring together: coloboma, heart defect, atresia choanae, retarded growth and development, genital hypoplasia, and ear anomalies/deafness.<sup>4</sup> Traditionally, CHARGE has been diagnosed if at least four of the six abnormalities are present. Several additional characteristics are commonly found in CHARGE patients. These include square face, asymmetrical facial features, broad nasal bridge, shallow orbital ridges, brainstem dysfunction resulting in feeding and respiratory problems, and cranial nerve immaturity.<sup>5,6</sup>

Coloboma is present in 80 to 90% of CHARGE patients.<sup>3</sup> Coloboma is an ophthalmic malformation that can occur in the iris, retina, optic nerve, or macula from an incomplete closure of the eye. Large colobomas of the retina and colobomas of the optic nerve or macula result in impaired vision.

Heart defects occur in 75 to 80% of CHARGE patients.<sup>3</sup> Conotruncal and aortic arch anomalies, particularly tetralogy of Fallot, are the most common heart defects seen in CHARGE patients.

Choanal atresia occurs in 50 to 60% of CHARGE patients.<sup>3</sup> In over 90% of cases of choanal atresia, the obstruction consists of bone as opposed to being membranous.<sup>7</sup> Bilateral choanal atresia poses an obvious problem to newborn infants. These patients display respiratory dis-

stress upon birth, resulting in cyanosis, which is temporarily relieved by crying.

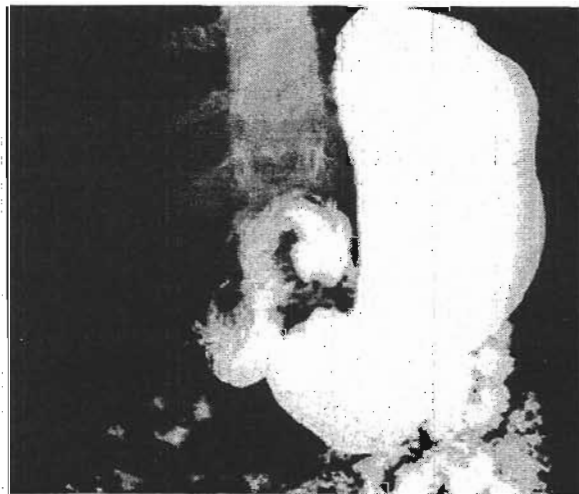
Children with CHARGE association commonly display developmental delay (100%) and growth deficiency (70%).<sup>3</sup> These patients have a wide range of mental capacity from normal intelligence to severe mental retardation.

Genital hypoplasia is found in both male and female CHARGE patients, occurring in 70 to 80% of cases. In females, hypoplasia of the labia and clitoris is observed; in males, micropenis and cryptorchidism are present. Treatment with testosterone may enlarge the penis to normal size and treatment with human chorionic gonadotropin may stimulate descent of the testes.<sup>3</sup>

About 90% of CHARGE patients display ear abnormalities, which can range from outer ear malformations to inner ear anomalies.<sup>3</sup> CHARGE patients often have laterally protruding ears, ossicular malformations, chronic serous otitis media, cochlear defects, and/or deafness.

Other characteristics manifested in CHARGE patients include velopharyngeal incoordination, facial palsy, cleft palate, tracheoesophageal fistula, esophageal atresia, and gastroesophageal reflux. Surgical intervention can alleviate some of the abnormalities associated with CHARGE, such as esophageal atresia or gastroesophageal reflux.

In this case, redundant duodenum and abnormal bands were the cause of the patient's gastrointestinal symptoms. Our patient had coloboma, developmental delay, growth deficiency, choanal atresia, genital hypoplasia, laterally protruding ears, and decreased hearing. As gastroesophageal reflux and vomiting are noted with CHARGE, we suggest that patients undergo radiologic examination



**FIG. 2.** Postoperatively, the duodenal redundancy is unchanged but there are no strictures present.

to ensure that the reflux is not caused by duodenal obstruction.

In our patient, congenital duodenal obstruction was diagnosed and relieved by laparoscopy. Laparoscopic diagnosis and treatment of rare congenital abdominal defects is feasible.

### ACKNOWLEDGMENT

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Address reprint requests to:  
*Constantine T. Frantzides, MD*  
*Evanston Northwestern*  
*2650 Ridge Avenue*  
*Barch 106*  
*Evanston, IL 60201*

*E-mail: cfrantzides@enh.org*