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# Should We Be Concerned About Jejunoileal Atresia During Repair of Duodenal Atresia?

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## **Abstract**

Introduction: During repair for duodenal atresia, it has been emphasized that inspection of the small bowel to identify a second atresia is required. The laparoscopic approach for repair of duodenal atresia has been criticized for its limitation to perform this step. Given that duodenal atresia and jejunoileal atresias do not share common embryologic origins, we question the validity of this concern. Therefore, we conducted a multicenter retrospective review of duodenal atresia patients to quantify the incidence of jejunoileal atresia in this population. Methods: After institutional review board approval (IRB #07-12-187X), a retrospective review was conducted on all patients who have undergone duodenal atresia repair at seven institutions over the past 7–12 years. Demographics and the presence or absence of a jejunoileal atresia were recorded.

**Results:** Four hundred eight patients with duodenal atresia were identified. The mean gestaational age was  $36.3 \pm 2.9$  weeks, and the mean weight was  $2.5 \pm 0.8$  kg. Mean age at operation was 19 days (range, 1–1314). There was a 28% incidence of trisomy 21. Two patients (0.5%) were identified as having a second intestinal atresia, and both were type IIIb. One patient was diagnosed at the time of duodenal atresia repair; the other was a delayed diagnosis. Both patients did well after repair.

*Conclusions:* In this, the largest series of duodenal atresia patients compiled to date, the rate of a concomitant jejunoileal atresia is less than 1%. This low incidence is not high enough to mandate extensive inspection of the entire bowel in these patients, and a second atresia should not be a concern during laparoscopic repair of duodenal atresia.

# Introduction

S EVERAL RESEARCHERS have suggested that patency of the entire gastrointestinal tract should be proven during repair of duodenal atresia. Therefore, the laparoscopic approach has been criticized for its limited ability to prove complete intestinal continuity. However, duodenal atresia and jejunoileal atresias do not share common embryologic origins. The incidence of concomitant jejunoileal atresia should, therefore, be unrelated to the presence of duodenal atresia. In order to evaluate this assumption, we conducted a multicenter retrospective review of duodenal atresia patients to quantify the incidence of jejunoileal atresia in patients with duodenal atresia.

# Methods

After institutional review board approval (IRB #07 12-187X), a retrospective review was conducted on all patients who have undergone duodenal atresia repair at seven institutions over the past 7–12 years. Demographics and the presence or absence of a jejunoileal atresia were recorded.

## Results

The compiled dataset included 408 patients with duodenal atresia. The mean gestational age was  $36.3\pm2.9$  weeks, and the mean weight was  $2.5\pm0.8$  kg. The mean age at operation was 19 days (range, 1–1314). The median age at operation was 3 days. There was a 28% incidence of trisomy 21.

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The duodenal atresia was repaired by the open approach in 348 patients and by laparoscopy in 60 patients. A second intestinal atresia was identified in 2 patients (0.5%), and both were type IIIb jejenoileal atresias. In 1 patient, the jejunoileal atresia was diagnosed at the time of open duodenal atresia repair, while the remaining patient was diagnosed several weeks after open repair by a contrast study that was performed to evaluate delayed return of bowel function. Both patients underwent primary anastomosis of the jejunoileal atresia with uneventful outcomes.

#### **Discussion**

Duodenal atresias and webs are believed to develop secondary to a low concentration of vacuole development after the solid cord phase of duodenal development from 8 to 10 weeks gestation, leading to failure of recannalization.<sup>6,7</sup> Jejunoileal atresias, on the other hand, are likely the result of vascular compromise of a section of intestine, resulting in obliteration of this segment, leaving two blind ends. The incidence of both lesions occurring in the same patient should, therefore, be extremely low and unrelated. In the largest series of duodenal atresia patients published in the United States, no cases of jejunoileal atresia were found in 169 consecutive cases.8 In another series of 187 patients from South Africa, there were no patients mentioned to have jejunoileal atresia. Incidence reports for jejunoileal atresia are widely variant, having been reported as high as 1 in 300, but is more conservatively estimated to occur in approximately 1 in 1000 live births. 10,11 If we apply these estimated occurrence rates to the duodenal atresia population, the 0.5% incidence found in our series, or 1 in 200, is only slightly outside the expected range. Combining our series with the other large series, the incidence would be 0.03%. Based on this incidence, the possibility for a partial link between conditions may exist, but the number of jejunoileal atresias found in this series of duodenal atresia patients is not high enough to rule out the possibility that they are an independent, occasionally overlapping phenomenon. Even in the presence of a concomitant distal atresia, the clinical relevance deserves further exploration.

While there are several researchers that recommend proving intestinal continuity during the repair of duodenal atresia, 1-5 there are several additional researchers who make no such suggestion. 9,12-16 This suggestion may be borrowed from experience with exploration for a known jejunoileal atresia where the possible presence of multiple atresias is well documented. 17,18 In addition to the low likelihood of an atresia beyond the duodenum, the ability to uncover it during the primary operation is limited by the proximal obstruction, which will result in decompression of all the distal bowel. Transition points with proximal dilation of a downstream atresia will not exist. Widely separated ends or apple-peeltype defects will usually be obvious whether the duodenal atresia is repaired open or laparoscopic. In this series, one such lesion was missed during an open duodenal atresia repair. Detecting distal webs or type 1 defects would require filling the entire small bowel with saline to prove patency, which is both a difficult and cumbersome step that risks bowel injury, and, according to the data we found, is probably not justified as a mandated operative step.

We routinely obtain an early contrast study between postoperative days 2–5 in our patients after laparoscopic duodenal atresia repair to prove patency, no leak, and gastric emptying, so we can remove the nasogastric tube and begin feeds regardless of the nasogastric output.<sup>19</sup> In a patient intolerant of oral feeds, or demonstrating delayed return of bowel function, this routine contrast study would likely reveal a distal obstruction at the point of jejunoileal atresia. Additionally, given that we currently repair jejunoileal atresias via an umbilical incision, a delayed diagnosis would not limit the benefits of minimally invasive surgery the patient enjoys after laparoscopic repair of the duodenal atresia.

#### Conclusions

In summary, there is no evidence that suggests that a search for a second intestinal atresia at the time of duodenal atresia repair is necessary, regardless of approach. Additionally, this report supports that the presence of a second atresia should not be a consideration when performing a laparoscopic repair of duodenal atresia.

### **Disclosure Statement**

No competing financial interests exist.

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