Characteristics and management of congenital esophageal stenosis: findings from a multicenter study.

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BACKGROUND: Congenital esophageal stenosis (CES) is a rare condition frequently associated with esophageal atresia (EA). There are limited data from small series about the presentation, treatment, and outcomes of CES.

METHODS: Medical records of all patients with CES included in the French Network on Esophageal Malformations and Congenital Diseases were reviewed retrospectively with regard to diagnosis, treatment, and outcome.

RESULTS: Over 18 years, 61 patients (30 boys) had CES, and 29 (47%) of these patients also had EA. The mean age at diagnosis was 24 months (1 day to 14 years) and was younger in patients with CES and EA than in those with isolated CES (7 vs. 126 months, p < 0.05). Twenty-one of the 61 patients with CES had no clinical symptoms: in three patients, the findings were incidental, and in 18 of the 29 patients with associated EA, CES was diagnosed at the time of surgical repair of EA or during a postoperative systematic esophageal barium study. In the 40 other patients, at diagnosis, 50% presented with dysphagia, 40% with vomiting, 50% with food impaction, and 42% with respiratory symptoms. Diagnosis of CES was confirmed by esophageal barium study (56/61) and/or esophageal endoscopy (50/61). Sixteen patients had tracheobronchial remnants (TBR), 40 had fibromuscular stenosis (FMS), and five had membrane stenosis (MS). Thirty-four patients (56%) were treated by dilation only (13/34 remained asymptomatic at follow-up); 15 patients were treated by dilation but required later surgery because of failure (4/15 remained asymptomatic at follow-up); and nine patients had a primary surgical intervention (4/9 were asymptomatic at follow-up). Dilation was complicated by esophageal perforation in two patients (3.4%). At follow-up, dysphagia remained in 36% (21/58) of patients, but the incidence did not differ between the EA and the isolated CS groups (10/29 vs. 7/32, p = 0.27).

CONCLUSIONS: CS diagnosis can be delayed when associated with EA. Dilation may be effective for treating patients with FMS and MS, but surgical repair is often required for those with TBR. Our results show clearly that, regardless of the therapeutic option, dysphagia occurs frequently, and patients with CES should be followed over the long term.

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