Double H-type tracheoesophageal fistula

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ABSTRACT

Congenital ‘isolated’ tracheoesophageal fistula (TEF) is a rare variant of esophageal atresia, also termed H-type TEF. Even rarer is a double H-type TEF. Here, we report a two-week-old female, with double H-type TEF – one identified pre-operatively, the other during surgery. Pre-operative laryngotraceobronchoscopy (LTB) with guidewire cannulation is considered key to definitive TEF repair, to aid in both pre- and intra-operative identification of the fistula(e). In our experience, only one H-type TEF was identified at the planned pre-operative LTB, with the second H-type fistula identified at the time of cervical dissection. Both fistulae were repaired during the same operation, via cervical approach. This represents the first reported case of a double H-type TEF, repaired primarily via a single cervical incision.

2. Introduction

Congenital ‘isolated’ tracheoesophageal fistula (TEF) without esophageal atresia (EA), also termed H-type TEF, is a rare variant of the EA spectrum. The H-type TEF has been reported to occur in approximately 3–4% of patients within the EA spectrum [1–4]. There have been very few cases reported in the literature of a double H-type TEF [5–12]. The first case reported in the literature was a post-mortem autopsy report by Hübner, in 1943 [11]. Since then, contemporary cases have reported double H-type TEF in survivors [5,6,8]. Here, we report a case of a two-week-old female, which represents the first reported case of a double H-type TEF, repaired primarily via a single cervical incision.

3. Case report

A two-week-old female was admitted to the neonatal intensive care unit for investigation of poor weight gain, noisy breathing after feeding, and episodes of cyanosis related to feeding. The patient was born via emergency low uterine segment caesarean section, indicated for fetal bradycardia. Her Apgar scores at one and five minutes, were nine and ten, respectively. Her gestational age was 39 + 2 weeks, and birth weight was 2.8 kg (3.5% centile, z score –1.8), as compared to her admission weight of 2.74 kg (2.8% centile, z score –1.9).

This patient was initially investigated with an upper gastrointestinal contrast study, which showed a possible but equivocal appearance for H-type TEF. She subsequently underwent a formal tube esophagogram, which did not confirm the finding of an H-type TEF. However, due to ongoing clinical suspicion, she was investigated further with laryngotracheobronchoscopy (LTB) to examine for a fistula. Again, initially, no H-type TEF was seen. However, following injection of methylene blue into the esophagus, a fistulous connection to the airway was identified and the presence of an H-type TEF confirmed.

On the day of H-type TEF repair, the neonate underwent LTB, at which a clearly visible H-type TEF was cannulated with a 0.32″ guidewire, prior to exploration via a right cervical (supravacular) approach. However, during cervical exploration, two distinct H-type TEF were identified and dissected. The more cranial TEF was encountered first and, notably, had no guidewire in situ. This TEF demonstrated the wider caliber of the two, and its identity was confirmed by passage of a 6Fr feeding tube into the esophagus with aspiration of gastric contents, as well as by a secondarily placed 0.28″ guidewire into the trachea. Image intensification was utilized to confirm that both the originally placed 0.32″ and secondarily placed 0.28″ guidewires were in situ (in the trachea cranially and esophagus caudally). The more caudal TEF was...
identified 8–10 mm caudally and contained the primarily inserted 0.32” guidewire. Both H-type TEF were divided and closed with interrupted 5/0 PDS (Polydioxanone) sutures. To discourage TEF recurrence, a strap muscle was mobilized, disconnected superiorly, and interposed between the trachea and esophagus. An intra-operative image is shown in Fig. 1A. Post-operatively, the patient was kept intubated, and returned to the neonatal intensive care unit.

Of note during the post-operative course was that extubation was delayed until 13 days post-repair due to suspected tracheal swelling. Prior to discharge from hospital on full breast feeds, the patient was screened for the VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, limb abnormalities) association. Whilst no VACTERL anomalies were identified per se, the patient was shown to have sensorineural hearing loss in the right ear.

Subsequent to discharge from hospital, the patient has had regular surgical and respiratory outpatient assessments. Over the ensuing four years, she has had no issues with feeding, gastroesophageal reflux, nor esophageal stricture. A contrast study was performed at three years of age, in the setting of new symptoms of intermittent stridor and upper airway secretions. This showed an irregular esophageal mucosal appearance at the level of the repair, albeit with no evidence of a recurrent TEF (Fig. 1B). Flexible upper gastrointestinal endoscopy affirmed the irregular mucosal appearance to represent the two sites of TEF closures, separated by an intervening bridge of tissue (Fig. 1C). Esophageal high resolution impedance manometry studies (Laborie/MMS) were performed at two and three years of age, as part of a larger study of esophageal dysmotility in EA patients, rather than for dysphagia or other symptom indications. The studies were analyzed using Swallow Gateway™ (swallowgateway.com; version © 2020), an online application [13]. Both studies demonstrated normal esophageal peristalsis (Fig. 2). Of note in the initial study, there was continuous increased pressure at the upper esophageal sphincter (not related to mechanical narrowing). However, at the second study 12 months later, there were no abnormally increased upper esophageal sphincter pressures. Now aged four years, the patient remains a healthy and thriving child, without requirements for gastroesophageal reflux management nor esophageal dilatation.

4. Discussion

Double H-type TEF was first reported in a post-mortem autopsy report by Hübner in 1943 [11]. Similarly post-mortem findings of double H-type TEF were reported by Leven et al. in 1952 [12], and Babbitt in 1957 [7]. In more recent times, Schulte et al. reported a case of a twenty-day-old male diagnosed with H-type TEF on contrast esophagogram, who underwent subsequent surgical correction of a single fistula [6]. Post-operatively, this patient had respiratory deterioration, and further contrast esophagogram demonstrated a likely
second H-type TEF. This was confirmed on rigid bronchoscopy, and the patient had a second operation to repair the second H-type TEF prior to discharge from hospital [6].

Only two previous reports have described patients with double H-type TEF, who underwent surgical repair of both TEF during the same operation. Mattei [5] reported a newborn male who had two TEF identified at the time of LTB. This patient required two incisions, one right posterolateral muscle-sparing thoracotomy incision, and one right cervical incision. De Carolis et al. [8] reported a three-day-old female who had two H-type TEF identified at bronchoscopy, but the operative management of the patient is not described. In our patient, although LTB only identified one H-type TEF, a second TEF was identified at the time of the cervical dissection, and both H-type TEF were repaired via the same right cervical incision.

5. Conclusion

The presence of double H-type TEF is a very rare clinical entity. Pre-operative LTB with guidewire cannulation is considered key to definitive TEF repair, to aid in both pre- and intra-operative identification of the fistula(e). In our experience, only one H-type TEF was identified at the planned pre-operative LTB, with the second H-type fistula identified at the time of cervical dissection. This notwithstanding, the lack of the guidewire within the first-encountered H-type TEF provided basis for further dissection and delineation of the second H-type TEF during the same operation. To the best of our knowledge, this is the first report that describes the repair of double H-type TEF during the same operation, via the same incision.

Patient consent

Informed consent regarding this case report has been obtained for the purpose of publication.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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