Congenital Esophageal Stenosis Associated with Esophageal Atresia

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Introduction

CES is suspected by a fixed intrinsic narrowing of the esophagus present at birth and associated with congenital malformation of the esophageal wall architecture [1]. Excluding the membranous type (MD), the diagnosis of CES is only confirmed by the histologic picture [2]. The histopathologic picture may show fibromuscular disease (FMD) or tracheobronchial remnants (TBR). The latter involves ciliated pseudostratified columnar epithelium, seromucous glands, or cartilage alone or in combination [3]. The association of CES and esophageal atresia (EA) and/or tracheoesophageal fistula (TEF) ranges from 0.4 % [1] to 14 % [4–6]. The authors believe that this association is common and that many cases are over looked. CES has been most frequently associated with EA with distal TEF (64 %), followed by isolated TEF (20 %) and isolated EA (16 %) [7]. In the authors experience, the incidence of CES being associated with pure atresia is higher than in EA and distal TEF (50 % vs 11.3 %) [6].

The first case of CES associated with an EA was reported from the Montreal Children Hospital by Dunbar in 1958 [8]. This was followed by 34 reports in the literature till 2008 [1, 4–7, 9–37]. Reviewing the histologic structure and the etiology of esophageal dysmotility in this association is mandatory for understanding this topic.

Histology of the Atretic Esophagus

Few reports are available in the literature regarding the histology of the atretic esophagus. Hokama et al. in 1986 [3] examined six autopsy unoperated cases of EA and TEF and also two surgical specimens. They defined the lower segment of the esophagus as that part where the wall is arranged into four normal layers: mucosa, submucosa, muscularis externa with myenteric plexus, and adventitia. The fistula was defined as that portion between the tracheal/bronchial connection and the transition to esophagus with normal layers. Tracheobronchial elements were defined as ciliated pseudostratified columnar epithelium, seromucous glands (as opposed to normal esophageal mucus glands), or cartilage, alone or in combination. Hokama et al. found a high incidence of TBR in the lower esophagus (five autopsy cases out of six and in the two surgical specimens). They concluded that TBR may be very common in EA/TEF that may lead to stenosis and abnormal motility after successful

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anastomosis. They also proposed that lack of a normal muscle coat at the fistulous end may cause esophageal dysmotility. They questioned the feasibility of including the fistula in the anastomosis should this correlation be confirmed.

In 1997, Merei et al. [38] from Australia carried out a histologic study of EA and TEF in an adriamycin animal model. They found that all the fistulae were lined with ciliated respiratory epithelium extending to a variable distances from the origin and in some instances as far as the stomach. Cartilage was occasionally seen in the wall. Transition from ciliated epithelium to stratified squamous epithelium occurred either by partial or abrupt replacement. The muscle layer was absent at the fistulous origin. Later, it was composed of irregular smooth muscle fibers that were not properly arranged into normal esophageal layers. After transition to normal esophageal epithelium, it became regular.

Dutta et al. in 2000 [39] reported a histological study of EA/TEF in 65 cases. The lining epithelium was stratified squamous in 36 cases, pseudostratified squamous in 2, and not seen in 27 cases. The mucous glands were abnormally high in number in 23 cases and with abnormal mucin secretion (typical of respiratory glands) in 23 cases. The ducts were dilated in six cases but with increased number in four. Cartilage was seen in eight cases with large number of mucous and seromucous glands also with abnormal mucin secretion. The muscularis propria was poorly oriented in 17 cases, well developed in 17, and disorganized in 13. Out of the studied cases, one autopsy case showed cartilage with mucous and seromucous glands. In another autopsy case, only mucous and seromucous glands were seen without cartilage. Both autopsy cases showed abnormal mucin secretion. The authors proposed that the TBR in the repaired esophagus as well as a disorganized muscle coat may be part of the transition from the fistula to a normal esophagus. The extent of this TBR is variable, and it may be premature to suggest this as a cause for esophageal dysmotility in each case. The authors stressed the point that loss of normal esophageal function may be due to abnormal numbers of glands and ducts and presence of abnormal mucin production. TBR may present with esophageal stricture refractory to dilatations but have dramatic response to resection. They claimed that these strictures are present only in the lower esophagus away from the area of anastomosis. However, this statement has been challenged. CES should be considered in the etiology of anastomotic stricture [6]. In our series, surgical specimens for histopathologic studies were obtained from the tip of the lower esophageal pouch during primary repair of EA/TEF cases [6]. Up to date, 10 patients out of 65 (15.4%) had histologic pictures suggestive of CES. None of the patients studied had absent muscle layers. This excludes using the fistula in the anastomosis. Two cases had fibromuscular disease (FMD), five with tracheobronchial remnants (TBR) without cartilage, and three with cartilage. The epithelium for these ten patients was normal in seven patients and pseudostratified columnar ciliated in three. These three cases showed increased numbers of mixed respiratory glands, ducts, and cartilage that extended from the submucosa to the adventitia causing muscle distortion (Fig. 9.1). One case with pure EA and gastric pull up showed TBR involving the whole lower esophageal pouch down to the cardia. Five patients showed mixed respiratory glands without cartilage that extended from the submucosa to the adventitia causing muscle distortion (Fig. 9.2). The glands were considered abnormal if they are seromucous or mucous glands that are increased in number and/or abnormally located outside the submucosa. The remaining two cases showed muscular hypertrophy and extensive fibrosis consistent with FMD (Figs. 9.3 and 9.4).

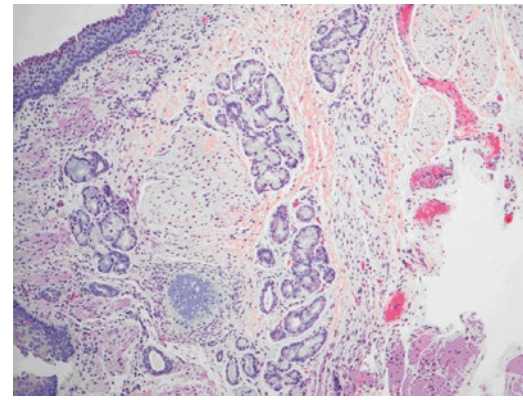

Fig. 9.1 Tracheobronchial remnants represented by pseudostratified columnar ciliated epithelium together with seromucous glands and cartilage (case 10)

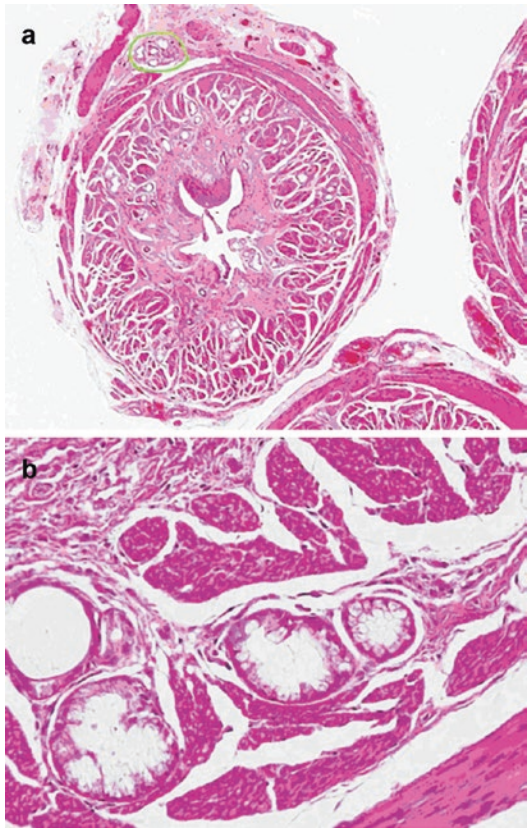

Fig. 9.2 (**a**) Mixed respiratory glands extending from the submucosa to the adventitia of the esophagus causing muscle disruption. (**b**) A magnified photograph of **a.** For the same patient

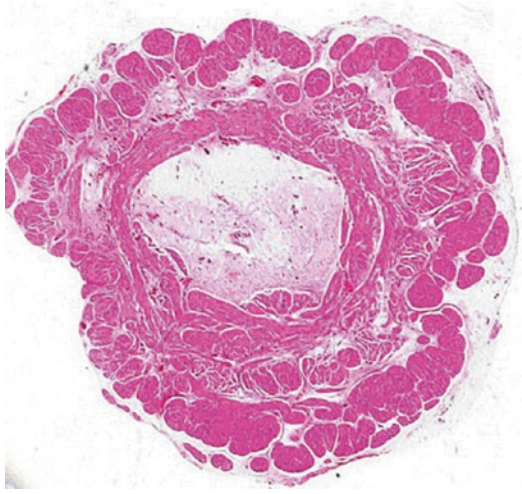

Fig. 9.3 Hypertrophic muscle fibers together with fibrosis

Etiology of Esophageal Motility Disorders in Esophageal Atresia

The etiology of esophageal dysfunction in cases of CES after repair of EA is not understood. It may be due to either CES or EA alone or in combination. *An acquired origin* for esophageal dysmotility is proposed. Extensive mobilization and denervation of the esophageal segments could aggravate reflux and motility disorders [40]. Normal peristaltic activity was documented preoperatively in the proximal esophagus in two patients who had EA without a fistula. One patient examined postoperatively showed a disturbed motility pattern [41]. Vagal damage may be the cause of motility dysfunction [42]. Extensive pouch mobilization is associated with severe motor disability [43].

A congenital origin for esophageal motility disorders after EA repair was proposed. Romeo et al. in 1987 [44] has documented disturbed motility preoperatively in patients with EA. Furthermore, motility disorder has been documented in patients having isolated TEF without EA [45]. Transection and anastomosis of the esophagus did not cause motility disorders [46]. Few histological studies have been conducted in the literature to document the congenital origin of esophageal dysmotility after successful repair of EA. Most of these studies have been done on autopsy patients [3, 47] or animal models [38, 48, 49]. Abnormal Auerbach's plexus was found in the esophagus and stomach in five autopsy patients with EA and TEF. The plexus was looser than normal in the distal esophagus and to a lesser extent in the proximal esophagus and stomach fundus. The ganglia were larger than normal. The smooth muscle layers were documented to be normal [47]. In other studies [3, 6, 38, 39], tracheobronchial elements, namely, ciliated pseudostratified columnar epithelium and seromucous glands with or without cartilage together with irregular smooth muscle fibers, were seen in sections of the distal esophagus. Seromucous glands were seen among muscle bundles of the lower esophageal pouch. Abnormal mucus glands causing muscle distortion were also documented. The transition from the fistula

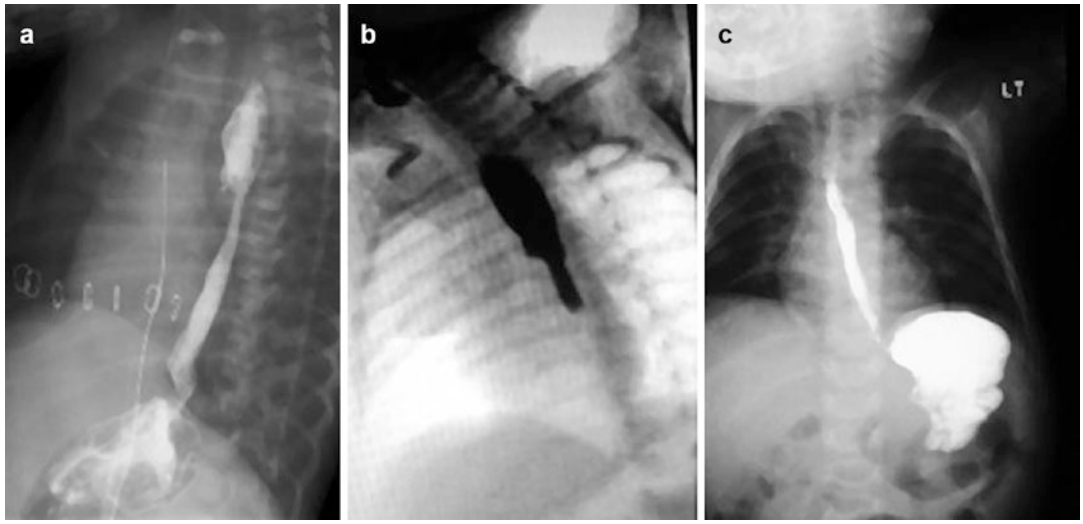

Fig. 9.4 Case 10 showing: (**a**) Initial normal barium swallow and meal. (**b**) Photo from videofluoroscopy with major dysmotility after 1 month with failure of passage of the contrast distally for more than 5 min. (**c**) Barium swallow and meal after fundoplication and lower esophageal myectomy showing improvement of motility

to normal esophageal histology takes place at a variable distance from the origin of the fistula and may extend down to the cardiac end [3, 6, 38]. Surgical specimen from the tip of the lower pouch also showed a histologic picture consistent with FMD [6].

Singarm et al. in 1995 [50] examined the histological and immunohistochemical features of CES of the FMD in two adults. In comparison to three controls, CES esophagi showed infiltration of neutrophils in the myenteric plane without any increase in collagen. NADPH diaphorase histochemistry showed a significant reduction of myenteric nitrinergic neurons and fibers of the circular muscle. The specific total lack of nitric oxide (NO) inhibitory innervation may be an important mechanism in the pathogenesis of stenosis and aperistalsis of the esophagus in this disorder.

Neuropeptides are abnormal in the atretic esophagus in the Adriamycin fetal rat model [48, 49] and in humans [51–53]. The density of the nerve plexus, ganglia, and number of cell bodies per ganglia immunostained by neuron-specific enolase (NSE), vasoactive intestinal peptide (VIP), or substance P (SP) was significantly reduced in EA/TEF fetuses. So, there are significant abnormalities of the intramural nervous components of the esophagus in EA/TEF fetal rats, involving both the excitatory (SP-labeled) and inhibitory (VIP-labeled) intramural nerves which may be the cause of esophageal dysmotility in EA/TEF [49].

The circumferential neuronal distribution in the myenteric plexus in the atretic esophagus of the rat model was reduced by 50%. The near-complete ring of nerve tissue along the plane of the myenteric plexus was replaced by clusters of nerve tissue in the atretic esophagus. This abnormal distribution of nerve tissue in the atretic esophagus may be contributing factor in the esophageal dysmotility seen in EA [54]. Qi et al. using the same model showed that the vagus nerve gave rise to fewer branches in the esophagus and assumed abnormal route at the level of the lower esophagus [55]. Boleken et al. examined the distal end of the proximal esophageal atretic segment of neonates undergoing EA/TEF repair for intrinsic neuronal innervation. They found that the distribution of ganglion cells and some nerve fibers is deficient. The inadequate and abnormal neuronal innervation of the esophagus could be related to esophageal dysmotility in EA. Deficient expression of glial cell line-derived neurotrophic factor (GDNF) could have an important role in the defective and/or abnor-

mal neuronal innervation of the atretic esophageal segment [51]. Similarly, Li Kai et al. in 2007 investigated the structural characteristics and the expression of a group of neuropeptides in the specimens obtained from the fistulous end of the lower esophagus of patients with EA/TEF. They found imbalance of neurotransmitters excretion in nerve vesicles, abnormal intrinsic dysplasia of nerve plexus, and increased expression of certain neuropeptides, e.g., VIP and nitric oxide synthase (NOS) were the main characteristics of the esophagus with abnormal intrinsic innervation, which may be responsible for postoperative esophageal dysfunction [52]. Most recently, Pederiva et al. examined the intrinsic esophageal innervation in children with isolated EA using specimens from the proximal and distal esophageal segments. There were denser fibrilar network and larger ganglia than controls [53].

Kawahara et al. in 2003 [56] investigated the motor function in four cases with isolated CES. Esophagogram showed stasis of contrast proximal to esophageal narrowing in two cases with FMD and one case with TBR. Three patients showed pathologic acid exposure by pH monitoring despite absence of evidence of esophagitis by endoscopy. Manometry showed synchronous esophageal contractions in FMD and TBR cases. LES pressure was at least 20 mmHg. Swallow-induced LES relaxation was incomplete in these cases. The authors concluded that gastroesophageal reflux (GER) and impaired esophageal motility are common in CES with FMD and TBR. Synchronous contractions seen in patients with FMD and TBR could be related to abnormal innervation of NO. The manometric data in CES in children reported by the Pittsburgh group showed segmental aperistaltic zone at the level of stenosis with local decreased pliability. The superior and inferior sphincters responded normally to swallowing [57]. A manometric study in 12 cases of CES showed another high-pressure zone (HPZ) in addition to the lower esophageal sphincter (LES) found in nine of the patients. This additional pressure zone disappeared after treatment [58]. Cheng in 2004 reported a case of achalasia-like esophageal dysmotility in a 14-year-old boy after successful repair of EA/TEF in the neonatal period. This was proved by clinical, radiological, and manometric study. The manometric features were a cardiac sphincter of 2 cm in length with a high pressure above 40 mmHg with aperistalsis in the esophageal body and failure of the cardiac sphincter to relax after a swallow. The condition responded well to Heller myotomy [59].

Kawahara in 2004 [60] reported the usefulness of videomanometry for studying pediatric esophageal motor disease in four postoperative cases of EA/TEF and one case of isolated distal CES due to TBR. These cases frequently showed impaired esophageal transit during defective esophageal peristaltic contractions. Videofluoroscopic image in cases of EA showed marked stasis of contrast in the esophageal body. Manometry showed absent contractions in the middle esophagus. The distal esophagus showed low-amplitude peristaltic contractions in two cases, low-amplitude synchronous contractions in one case, and no contractions in one case. The authors concluded that impaired esophageal transit was caused by defective luminal closure especially in the middle esophagus during deglutition, but not by LES malfunction. Videofluoroscopy of the case of CES showed stasis of the contrast in the distal dilated esophagus associated with narrowing at the end of the esophagus mimicking achalasia. Manometry showed swallow-induced LES relaxation and low-amplitude synchronous contractions in the whole esophageal body.

Types of Congenital Esophageal Stenosis

Fekete et al. [1] defined CES as intrinsic stenosis caused by congenital malformation of the esophageal wall. It involves a type with TBR, another with segmental hypertrophy of the muscularis and diffuse fibrosis of the submucosa (FMD), and a third type with a membranous diaphragm (MD). The most common type of CES was that of the TBR variety (75 %) followed by the FMD (25 %) [4, 6]. These percentages might not be correct. The FMD responds better to balloon dilatation. If dilatation is successful, no specimen is available. So, a definitive subtype cannot be

determined. Also the percentage of each type that will respond to dilatation is not known [34]. The MD type is rarely reported in association with EA/TEF [9, 32, 61]. It is possible that CES can be multiple [62]. The stenotic area may involve the perianastomotic area or even may extend distally to a variable distance [4, 6]. For this reason, we disagree with the statement that CES does not involve the anastomotic site and is always separate from it [1, 35]. However, a distal isolated area of CES separate from the anastomotic site may be present [4, 5, 7, 15]. All the 11 cases reported by Kawahara in 2001 were found to have narrowing between the anastomosis and the gastroesophageal junction: in the mid-esophagus in two and in the lower esophagus in nine patients [4]. In our experience with ten cases, only one of them had distal esophageal stricture due to FMD that required resection. So, in our series, CES is more common at the perianastomotic area (Tables 9.1, 9.2, and 9.3).

Diagnosis

A high index of suspicion should be raised in all cases of EA. CES is an intrinsic stenosis that may be present at birth but not necessarily symptomatic [7]. The diagnosis of CES is suspected in a neonate with EA if a size 8 French nasogastric tube cannot be passed into the stomach [4, 7]. However, passage of the tube down to the stomach does not rule out CES [7]. The diagnosis of CES before or during primary repair of EA is possible; however, simultaneous repair of the stenosis by doing a double esophageal anastomosis is a controversial approach [4, 32]. An esophagogram demonstrating a narrow segment above the cardia is radiologically diagnostic for CES when found in the neonatal period [7, 63]. Minor esophageal dysmotility as detected by barium is defined as aperistalsis, antiperistalsis, and simultaneous or uncoordinated contractions. The dysmotility is considered major if the transit time for

Table 9.1 Group I (two cases with FMD)

Criteria	Case 1: EA/TEF	Case 2: EA/TEF
Sex	Female	Female
GA/BW	33 weeks/1.9 kg	37 weeks/2.9 kg
Histopathology[a]	Unremarkable	FMD
Initial barium	No dysmotility, GER++, no stricture	No dysmotility, GER ++, no anastomotic stricture
Early postoperative period	Uneventful	Uneventful
Onset of symptoms	3 months, mainly dysphagia, aspiration, and FTT	2 months, mainly dysphagia, aspiration, and FTT
Subsequent barium	Anastomotic stricture extending distally	Anastomotic stricture
	GER +++	GER +++
	Major lower esophageal dysmotility	Minor dysmotility
Esophagoscopy	Normal mucosa	2nd degree esophagitis
Action	Failed antireflux medical treatment and dilatation	Failed medical antireflux measures and frequent dilatation
	Failed myotomy/Nissen fundoplication with gastrostomy at 6 months	Nissen fundoplication at 6 months
	Resection of distal stenotic area at 9 months showed FMD	Esophageal diverticulectomy at 14 months
		Required 4 dilatations
Outcome	Improved	Improved
Follow-up period	12 years	10 years

GA gestational age, *BW* birth weight, *FTT* failure to thrive
[a]Histopathology = surgical specimen from the tip of L.P at primary or delayed primary repair

Table 9.2 Group II (five cases). TBR without cartilage

Criteria	Cases 3, 4, and 5: EA/TEF	Case 6: pure EA (delayed repair at 5 months)	Case 7: EA/TEF
Sex	Female	Female	Female
	Male		
	Female		
GA/BW	33 weeks/1.8 kg	37 weeks/2.4 kg	37 weeks/2.5 kg
	37 weeks/2.4 kg		
	35 weeks/1.7 kg		
Histopathology	TBR/no cartilage	TBR/no cartilage	Operated somewhere else/no pathology specimen
Initial barium	No dysmotility in one, minor dysmotility in 2, GER ++ in all, slight stricture in all	Minor dysmotility	Minor dysmotility
		GER ++	GER ++
		No stricture	No stricture
Early postoperative period	Uneventful	Uneventful	Uneventful
Onset of symptoms	2 months, mainly slow feeding and occasional aspiration	10 months mainly dysphagia, FTT, and recurrent aspiration	3 months, dysphagia, aspiration, FTT
Subsequent barium	Isolated anastomotic stricture	No stricture	Stricture +++
	Minor dysmotility	Minor dysmotility	Minor dysmotility
	GER +++	GER +++	GER ++
Esophagoscopy	Norma mucosa in all	2nd degree esophagitis	Normal
Action	Medical antireflux measures and dilatation	Medical antireflux measures failed Thal's fundoplication and temporary gastrostomy	Failed medical antireflux measures Frequent dilatations failed Thal's/gastrostomy Failed dilatation Anastomotic resection → TBR without cartilage
Follow-up period/ outcome	5–9 years	7 years	3 years
	Improved	Improved	Improved

the bolus to go to the stomach is greater than 5 min [6]. The problem of major dysmotility is that it develops late and can be fatal or amenable to major complications.

Taking a surgical specimen routinely from the tip of the lower pouch during primary repair of EA may show a histological picture consistent with FMD or TBR [6]. This mandates close observation. The latest case in our series (case 10) showed TBR with cartilage (Fig. 9.1). The initial esophagogram was normal. A repeat fluoroscopic esophagogram 1 month later showed major esophageal dysmotility due to aperistalsis in the perianastomotic area. The patient required a feeding gastrostomy, anterior partial fundoplication, and lower esophageal anterior myectomy. The major esophageal dysmotility as seen at fluoroscopy improved, and partial oral feeding was allowed. The histology of the myectomy was normal. We learn from this case that the histologic picture confirmed the site and type of CES. The forthcoming scenario was anticipated. Dysphagia which developed after 1 month was not due to introduction of solid food but was due to pure dysmotility which preceded mechanical stricture. The lower esophageal myectomy helped to improve major dysmotility probably due to a decrease in the pressure of the lower high-pressure zone.

The late-onset diagnosis is suspected by the clinical triad of recurrent aspiration, dysphagia, and FTT together with the aid of an esophago-

Table 9.3 Group III. TBR with cartilage (three cases)

Criteria	Case 8: EA/TEF	Case 9: pure EA	Case 10: EA/TEF
Sex	Female	Male	Male
GA/BW	30 weeks/1.3 kg	35 weeks/2 kg	37/2.5 kg
Histopathology	TBR with cartilage	TBR with cartilage	TBR with cartilage
Initial barium	Minor dysmotility, GER ++ Slight anastomotic stricture	–	Normal
Early post-operative period	Uneventful	–	Uneventful
Onset of symptoms	3 months, dysphagia, aspiration and FTT	–	One month/slow feeding
Subsequent barium	Stricture at anastomotic site extending distally Late major dysmotility GER ++++	–	Major esophageal dysmotility at 1 month No GER
Esophagoscopy	Scope could not pass	–	–
Action	Medical antireflux measures and dilatations failed Thal's fundoplication and gastrostomy at 6 months Recurrent symptoms Resection of anastomotic stricture at 1 year. *Histopathology*: TBR with cartilage Recurrent stricture Frequent dilatations	Failed delayed primary repair Required resection of the whole lower esophagus and gastric pull-up *Histopathology* showed TBR with cartilage extending from the tip down to cardiac end of the lower esophagus	Thal's fundoplication Gastrostomy at 6 weeks Lower esophageal myectomy showed normal histology Partial oral feeding and gastrostomy
Follow-up/outcome	Improved now 6 years old		Improved now 9 months old

gram [64]. Patients may present with distal esophageal foreign bodies [7]. A barium study is the diagnostic and follow-up tool. Full cooperation between the radiologist and the surgeon is required.

The presence of GER in cases of EA with CES is said to be unlikely [27]. However, others believe that it is common [4–6]. Esophagoscopy and biopsy, pH monitoring, and possibly manometry may be required [58]. However, even with these investigations, it may be difficult to differentiate between CES and stricture due to GER [7]. Errors in diagnosis are common as most of these patients are diagnosed and managed as peptic stricture [5, 7].

Precise preoperative diagnosis of CES is important. The type of stenosis determines the modality of treatment. Without the preliminary histologic picture, the preoperative differentiation between FMD and TBR with cartilage is difficult. In a review of the literature for 59 cases with TBR with cartilage up to 2004, a correct preoperative diagnosis of the underlying etiology of stenosis was not reached in most cases. The majority were diagnosed as achalasia or peptic stricture [64]. Esophagoscopy and biopsy may fail to show deep-seated ectopic tissue [1, 65]. Fluoroscopy may show abrupt narrowing in cases of TBR, while that of the FMD may show more gradual, regular, and well-centered narrowing. However, fluoroscopy does not always show these typical findings [35]. During balloon dilatation, the presence of a short, sharp waist-like impression which suddenly disappears with increased pressure means the presence of cartilaginous rings [7, 34]. Endoscopic ultrasonography is said to be useful to distinguish TBR with cartilage from FMD [35, 66, 67]. Intraoperative palpation and the use of the flexible esophagoscopy may be of help [35, 65]. The author like others [58] found that intraoperative palpation of the lesion is not always easy. However, these modali-

ties suspect but not confirm the presence and the type of CES. The diagnosis is only confirmed by the histologic picture.

Treatment

Cases of EA diagnosed or suspected to have associated CES in the neonatal period should have utmost attention. A normal early fluoroscopic barium swallow and meal does not exclude CES and must be repeated at 4–6 weeks. The development of the triad of esophageal dysmotility, GER, and stricture should be managed as early as possible. By doing this, the consequences of malnutrition, recurrent aspiration, and even mortality can be avoided. Full antireflux measures and balloon dilatation for stricture should be initiated. If the dysmotility is major, a partial anterior wrap together with a feeding gastrostomy is indicated. During the procedure, a myectomy as long as possible of the lengthened lower esophagus is taken and sent for histopathologic and possibly histochemical examination.

For those cases discovered late, proper chest treatment, antireflux measures, and nutritional support should be started. Antireflux measures together with balloon dilatation should be the initial treatment of all forms of CES associated with EA [7, 34].

Most cases with TBR without cartilage respond well to medical antireflux measures together with balloon dilatations. Three out of four patients in our series showed excellent long-term clinical outcome despite the persistence of radiological minor dysmotility. Only one patient required surgical resection after failed balloon dilatations. Histopathology of the resected specimen showed glands without cartilage.

Cases with TBR with cartilage will require a limited surgical resection and primary anastomosis if balloon dilatations failed on three occasions [4, 5, 7, 35]. Surgical resection will also be indicated if initial sufficient dilatation is not achieved or symptoms recur very soon after dilatation [58]. Unnecessary prolonged trials of balloon dilatations should be avoided because the trials will be unsuccessful [64]. Some authors experienced severe complications with repeated dilatations [4, 68, 69]. The extent of TBR into the distal esophagus should be accurately assessed during surgery. A frozen section biopsy may be required. A fundoplication is recommended after resection if it disturbs the gastroesophageal junction and to avoid the possible postoperative complications of GER and hiatus hernia [1, 7, 33]. Circular myectomy was performed successfully for the treatment of CES due to TBR [70, 71]. Thoracoscopic resection of a distal CES and esophageal end to end anastomosis was successfully performed [72]. Very recently, a laparoscopic lower esophageal stricturoplasty with anterior fundoplication for CES due to TBR was also successful [73]. Surgical resection may be complicated by recurrent anastomotic stricture that may require few postoperative dilatations.

Cases with FMD usually respond to balloon dilatations [4, 7, 35]. Longitudinal myotomy with Nissen fundoplication may be curative [4, 35]. A limited surgical resection may be required if the above measures fail [4, 35, 58].

Cases of MD and multiple stenoses can be treated with esophageal dilatations alone. Endoscopic partial resection of the membrane can be done at the time of dilatation [35].

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