Summary and Conclusions

- CES is rare but common if associated with EA.
- The etiology of esophageal dysfunction in CES is complex. It can be due to CES itself or associated EA or both in combination. It includes textural abnormalities, excessive dissection, GER and the development of stricture. The muscles may be hypoplastic, Hypertrophic, distorted by fibrosis, cartilage and/or respiratory glands. There are abnormalities in the intrinsic and extrinsic innervation of both esophageal pouches. Neuropeptides are also abnormal. Lack of nitric oxide (NO) inhibitory innervation may be an important mechanism in the pathogenesis of stenosis and dysmotility. Also, there are ultrastructural abnormalities in EA.
- The present definition of CES proposed by Fékété is excellent for the intrinsic intramural part of the disease. The definition needs to be revised to include the spectrum of CES.
- CES can affect the anastomotic site of EA, extends distally to the cardia or be a separate distal CES away from the anastomosis and cardia.
- IF the CES involves the GEJ, it behaves like achalasia cardia with diagnostic challenges.
- CES is a spectrum of diseases that can behave differently according to the type, site and severity.
- The true incidence of CES is not known due to difficulties in diagnosis, successful dilatation and failure to obtain a histological diagnosis.
- There are difficulties in diagnosing CES in neonates due to absence of histology specimens and lack of a high index of suspicion during initial esophagogram after primary repair.
- Lack of miniprobe endoscopic ultrasonography (MEUS) facility and expertise adds to the diagnostic difficulties of CES.
- The presentation of CES can be acute and stormy in the neonatal period or it can be delayed most commonly to the weaning period. The course of the disease can be benign to present in late infancy, childhood or even in adults.
- CES can be a risk factor for anastomotic leak, recurrent TEF, refractory and recurrent esophageal stricture.

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- Neonatal diagnosis of CES can be improved by a routine passage of a transanastomotic size 8 NGT during primary repair, obtaining biopsies from the tips of esophageal pouches for histology and having a high index of suspicion during initial esophagogram. Anastomotic leak or recurrent TEF can be a shadow for a distal CES.
- Intolerance to feeds, recurrent respiratory symptoms (apnea, desaturations, aspiration, and choking…) are the main symptoms in neonates and young infants. Foreign body impaction and dysphagia are the main symptoms in older children.
- The triad of stenosis, dysmotility and GER is not uncommon in CES associated with EA sparing the GEJ.
- Successful NGT feeding is a good test to diagnose obstruction at the esophageal level.
- Esophagogram (with a low threshold for repetition), PH, impedance studies, upper endoscopy, esophageal manometry, and EUS are important tools to differentiate CES from cardiac achalasia and reflux esophagitis.
- Intra operative palpation for CES and/or frozen section biopsies are helpful in doubtful cases of CES if EUS is not available.
- The initial line of treatment is always dilatation whatever the type of CES is. The type of stenosis then determines the modality of treatment as diagnosed by EUS to achieve successful results without undue prolonged courses of dilatations and complications.
- Many cases diagnosed as neonatal or infantile cardiac achalasia are probably cases of CES that either require resection after failure of Heller's myotomy (TBR) or respond to dilatation or myotomy if it is FMD subtype.
- Isolated CES responds better to treatment than CES associated with EA.
- Gastrostomy and partial fundoplication may become mandatory should the triad of stenosis, dysmotility and GER develop.
- No heroic esophageal replacement surgery except after full chance of conservative management has been offered.

Recommendations

- Obtaining histopathologic samples from the tips of the esophageal pouches routinely during primary repair are helpful for early diagnosis of CES in the neonatal period.
- A transanastomotic size 8 Fr NGT is mandatory to rule out CES during primary.
- A high index of suspicion should be practiced during the initial esophagogram. Consider anastomotic leak and recurrent TEF as a possible shadow for a distal CES.
- A low threshold for repeating esophagogram is required if still in doubt.
- Early diagnosis and prophylactic dilatation should start as early as 4 weeks after primary repair of EA.
- The miniprobe EUS is helpful for the diagnosis of CES and its subtypes and the decision about the treatment modality and should be used in a wider scale.
- Protracted courses of dilatations with adjuncts, indwelling balloon dilatations, incisional therapy, and stents are preferable to esophageal replacement surgery despite of the possible complications which we hope to be minimized in the future with better techniques and better quality of stents.

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