Vascular Anomalies Associated With Esophageal Atresia and Tracheoesophageal Fistula: Incidence, clinical presentation, diagnosis and consequences

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Introduction: Congenital vascular anomalies have been shown to be associated with EA/TEF. Four types have been previously reported: 1- right aortic arch (RAA) and an aberrant left subclavian artery (ALSA); 2- RAA with mirror image; 3- left aortic arch and an aberrant right subclavian artery (ARSA); and 4- double aortic arch [1].

1- ALSA
2- RAA+ mirror
3- ARSA
4- Double aortic arch

Objective: To report their incidence in a cohort of patients while describing clinical presentation, diagnosis and consequences. The secondary objective is to evaluate the diagnostic value of esophogram in the diagnosis of ARSA.

Methods: All patients born with EA/TEF, from 1985 to 2013 were studied. Preoperative echocardiography reports, surgical description of primary esophageal repair and esophagograms were retrospectively reviewed. Age at diagnosis, discovery mode, clinical presentation and need for surgical correction of the vascular malformation were noted.

Results and Discussion:
- 18% had a vascular malformation. The incidence of RAA and ARSA was 6% (5/76) and 12% (9/76), respectively.
- Long gap EA and severe cardiac malformations requiring surgery were both significantly associated with vascular anomalies (p<0.05).
- Often asymptomatic, these abnormalities may be the cause of respiratory symptoms (dyspnea, cough, cyanosis) and/or exacerbate gastrointestinal symptoms (dysphagia) when a ring completely or incompletely encircles the trachea and/or the esophagus resulting in extrinsic compression [2].
- Defining the liability of the vascular malformation is often impractical in the setting of children operated on EA/TEF, because it may accompany various anomalies, including tracheomalacia, gastroesophageal reflux, anastomotic stricture, or congenital stenosis of the esophagus.
- Severe complications such as massive gastrointestinal bleeding secondary to an ARSA-esophageal fistula have also been reported [3].
- 254 esophagograms were reviewed; 40% were inconclusive for the detection of vascular anomalies. The diagnosis of vascular malformation was missed in four patients with a long gap EA. The sensitivity of esophagogram for the diagnosis of ARSA was 66%, the specificity was 98%, the negative predictive value 95%, and the positive predictive value 85%.
- The physician's attention must be focused on the specific search of aortic arch anomalies, the procedure must be made under good conditions (must include not only oblique images) and no major postoperative alterations should impair the interpretation.

Surgical repair of vascular malformation is indicated when the vascular ring is complete and respiratory and/or digestive symptoms are attributable to the malformation, which is very rare in practice [4].

Conclusion: ARSA and RAA have an incidence of 12% and 6% respectively, in EA/TEF patients. Echocardiography and esophagogram are effective but their sensitivity is not optimal for the diagnosis of ARSA. A CT-angiogram is recommended to rule out such malformations when esophageal stenting is indicated.

References: