Vascular Anomalies Associated with Esophageal Atresia and Tracheoesophageal Fistula

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Objective To report the incidence of congenital vascular anomalies in a cohort of patients with esophageal atresia (EA) and tracheoesophageal fistula (TEF) while describing the clinical presentation, diagnosis, and consequences, and to evaluate the diagnostic value of esophagram in diagnosing an aberrant right subclavian artery (ARSA).

Methods All patients born with EA/TEF between 2005 and 2013 were studied. Preoperative echocardiography reports, surgical descriptions of primary esophageal repair, and esophagrams were reviewed retrospectively.

Results Of the 76 children born with EA/TEF included in this study, 14 (18%) had a vascular malformation. The incidence of a right aortic arch (RAA) was 6% (5 of 76), and that of an aberrant right subclavian artery (ARSA) was 12% (9 of 76). RAA was diagnosed in the neonatal period by echocardiography (4 of 5) or surgery (1 of 5), and ARSA was diagnosed by echocardiography (7 of 9) or later on the esophagram (2 of 9). Respiratory and/or digestive symptoms occurred in 9 of the 14 patients with vascular malformation. Both long-gap EA and severe cardiac malformations necessitating surgery were significantly associated with vascular anomalies ($P < .05$). The sensitivity of the esophagram for diagnosing ARSA was 66%, the specificity was 98%, the negative predictive value was 95%, and the positive predictive value was 85%.

Conclusion ARSA and RAA have an incidence of 12% and 6% respectively, in patients with EA/TEF. A computed tomography angioscan is recommended to rule out such malformations when stenting of the esophagus is indicated, before esophageal replacement surgery, and when prolonged (>2 weeks) use of a nasogastric tube is considered. *(J Pediatr 2015;166:1140-4)*.

Esophageal atresia (EA) and tracheoesophageal fistula (TEF) are frequent congenital malformations (1 in 2500 births), characterized by interrupted continuity of the esophagus with or without a persistent communication with the trachea.[1] Associated anomalies, including cardiac defects, renal anomalies, anal or duodenal atresia, and vertebral malformations, occur in 30%-50% of cases.[1] Congenital vascular anomalies also have been associated with EA/TEF. Four types of anomalies have been described: right aortic arch (RAA) and aberrant left subclavian artery (ALSA); RAA with mirror image; left aortic arch and aberrant right subclavian artery (ARSA); and double aortic arch.[2]

In a child with EA/TEF, the diagnosis of RAA is necessarily made during the neonatal period either by echocardiography or by the surgeon who may be hampered in a right thoracic approach. In contrast, ARSA is not visible during esophageal surgery and is difficult to diagnose. Often asymptomatic, these abnormalities may be the cause of respiratory symptoms (eg, dyspnea, cough, cyanosis) and/or may exacerbate gastrointestinal (GI) symptoms (dysphagia) when a ring completely or incompletely encircles the trachea and/or the esophagus, resulting in extrinsic compression. Severe complications, such as massive GI bleeding secondary to an ARSA–esophageal fistula, have been reported as well.[3]

To date, published large case series of EA/TEF have focused exclusively on RAA.[4-9] The main objective of the present study was to report the incidence of vascular anomalies in a cohort of EA/TEF while describing the clinical presentation, diagnosis, and consequences. In addition, we sought to evaluate the diagnostic value of the esophagram in diagnosing ARSA.

Methods

This study was a retrospective review of all patients born with EA/TEF between January 2005 and October 2013 and followed at Sainte Justine Hospital. The study was approved by the local Institutional Review Board.

ALSA	Aberrant left subclavian artery
ARSA	Aberrant right subclavian artery
EA	Esophageal atresia
GI	Gastrointestinal
MRI	Magnetic resonance imaging
RAA	Right aortic arch
TEF	Tracheoesophageal fistula
VACTERL	Vertebral, anorectal, cardiac, tracheal, esophageal, renal, limb

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Patients were excluded from the analysis if they had been operated on elsewhere or if death occurred in the first week of life. Data were collected using standardized data abstraction forms. The abstracted information included the following patient-level variables: sex, birth weight, type of atresia, cardiac and other anomalies, VACTERL association (association of at least 3 of the following malformations: vertebral, anorectal, cardiac, tracheal, esophageal, renal, or limb), associated syndromes, and chromosomal anomalies. It also included the following surgery-level variables: "long-gap," 1-step surgery, 2-step surgery, and age at surgery. A long gap between the 2 ends of the esophagus in the cases of EA was defined as a gap longer than 3 cm or greater than the height of 2 vertebral bodies.[10]

Subjects with EA/TEF and associated vascular anomalies were identified. The presence of an ARSA or RAA (mirror image, ALSA, or double aortic arch) was demonstrated by preoperative echocardiography report, surgical description of primary esophageal repair, or the presence of a posterior indentation on the proximal esophagus detected on an upper GI series. In our institution, preoperative echocardiography in children with EA is performed according to a specific protocol in which the position of the aortic arch (right or left) and the presence of an aberrant vessel are systematically reported. Age at diagnosis, discovery mode, clinical presentation, and need for surgical correction of the vascular malformation were noted.

To assess the diagnostic value of the esophagram in detecting ARSA or ALSA, all upper esophageal series were blindly reviewed. In the esophageal series, the presence of an aberrant vessel was defined by a posterior indentation (**Figure 1**; available at www.jpeds.com). An examination was considered inconclusive for the specific search of vascular anomalies if the images were oblique (as opposed to completely lateral), if the patient ingested little contrast, if the study was done in a retrograde manner via a gastrostomy, or if there was a leak image at the site of anastomosis, masking the posterior wall of the esophagus on the lateral view. Reviews were conducted alphabetically and blind to the presence or absence of vascular anomaly.

Statistical Analyses

All statistical analyses were performed with SAS version 9.0 (SAS Institute, Cary, North Carolina). The Student t test was applied to compare means, and the χ^2 test was used to compare percentages. $P < .05$ was considered to indicate statistical significance. The estimation of CIs was set at 95%. The diagnostic values (ie, sensitivity, specificity, positive predictive value, negative predictive value) were calculated for an upper GI series.

Results

A total of 86 patients with EA/TEF born between January 2005 and October 2013 were seen at Sainte Justine Hospital. Ten children had insufficient data or follow-up, including 3 children who died in the first week of life and 7 children who underwent surgery elsewhere (**Figure 2**; available at www.jpeds.com.)

Seventy-six patients (36 females; 47%) met the inclusion criteria. The median birth weight was 2530 g (range, 1065-4400 g), and median gestational age was 37 weeks (range, 29-41 weeks). Seven patients (9%) were born of a twin pregnancy, but each was the only sick child of the twin pair. Forty of the 76 children (52%) were born in another hospital and transferred at birth to Sainte Justine Hospital. A prenatal diagnosis of EA/TEF was made in 29 cases (38%).

According to the Gross classification scheme,[1] 60 patients (79%) had EA type C (distal fistula), 6 (8%) had type A (isolated atresia), 5 (7%) had type E (isolated fistula), 4 (5%) had type D (proximal and distal fistulas), and 1 (<1%) had type B (proximal fistula). The types of surgery performed were thoracotomy in 57 children, thoracoscopy in 14, and cervicotomy in 5.

A long-gap EA was present in 12 patients (15%), with the same proportion of type A and type C. VACTERL association was present in 18 patients (24%). Forty-two patients (55%) had a cardiac anomaly, including an atrial septal defect in 25, ventricular septal defect in 7, tetralogy of Fallot in 2, atrioventricular communication in 2, aortic stenosis in 2, aortic isthmus hypoplasia in 2, transposition of the great arteries in 1, and coarctation of the aorta in 1. Fewer than one-quarter of these patients (10 of 42) required cardiac surgery. Surgery was performed for ventricular septal defect, tetralogy of Fallot, and atrioventricular communication in 2 children each and for transposition of the great vessels, aortic stenosis, coarctation of the aorta, and ductus arteriosus in 1 child each.

A vascular malformation was found in 14 children (18%). Characteristics of the patients with vascular anomalies are summarized in **Table I**.

RAA

Five of the 76 patients (6%) had an RAA, including 2 with ALSA and 3 with mirror image. One of these 5 children with an RAA had a long-gap EA, and 4 (80%) had a cardiac malformation. Two children required surgery for tetralogy of Fallot. The diagnosis was suspected on the first day of life in all 5 patients, either based on echocardiography (n = 4) or during the thoracic approach by the surgeon (n = 1). An angioscan and/or magnetic resonance imaging (MRI) were performed in 3 patients, which confirmed and depicted the anatomy of the vascular malformation. Despite the discovery of an RAA, none of the 4 surgeons changed the side of initial thoracotomy or thoracoscopy.

Two children with an RAA and mirror image remained completely asymptomatic during follow-up. One child died of cardiac malformation despite surgery. Severe respiratory and/or digestive symptoms occurred in the 2 patients with an RAA and ALSA, despite treatment of comorbidities (eg, tracheomalacia, cardiac anomaly, anastomotic stricture). In these 2 cases, correction of the vascular ring resulted in a total resolution of symptoms. **Table II** (available at www.jpeds.

Table I. Characteristics of patients with vascular anomalies

Patient	Vascular anomaly	Age at diagnosis	Mode of diagnosis	Type of EA	VACTERL	Cardiac anomaly	Cardiac surgery
1	RAA + mirror	1 d	Echocardiography	B	No	ASD	No
2	RAA + mirror	1 d	Echocardiography	C	Yes	TOF	Yes
3	RAA + ALSA	1 d	Echocardiography	D	No	ASD	No
4	RAA + mirror	1 d	During surgery	C	No	-	No
5	RAA + ALSA	1 d	Echocardiography	C long gap	Yes	TOF	Yes
6	ARSA	1 d	Echocardiography	A long gap	Yes	CoA	Yes
7	ARSA	8 mo	Esophagram	C	No	-	No
8	ARSA	1 d	Echocardiography	C	Yes	AVSD	Yes
9	ARSA	1 d	Echocardiography	C long gap	No	AIH	No
10	ARSA	1 d	Echocardiography	C	No	Aortic stenosis	No
11	ARSA	1 d	Echocardiography	C	No	-	No
12	ARSA	1 d	Echocardiography	A long gap	Yes	VSD	Yes
13	ARSA	1 d	Echocardiography	C long gap	Yes	ASD	No
14	ARSA	6 y	Esophagram	E	No	-	No

AIH, aortic isthmus hypoplasia; *ASD*, atrial septal defect; *AVSD*, atrioventricular septal defect; *CoA*, coarctation of aorta; *TOF*, tetralogy of Fallot; *VSD*, ventricular septal defect.

com) summarizes the clinical presentation of each of the 5 children with an RAA.

Left Aortic Arch with ARSA

Nine of the 14 patients with vascular malformation had an ARSA, representing an incidence of 12% in our cohort. Four of 9 children with an ARSA (45%) had a long-gap EA, and 6 of these 9 children (66%) had a cardiac malformation. Three cases required surgery in the neonatal period for coarctation of the aorta, atrioventricular communication, and ventricular septal defect. The majority of ARSAs (7 of 9) were diagnosed by preoperative cardiac ultrasound. In 2 children, the diagnosis was performed after barium swallow, one at age 8 months and the other at age 6.5 years. The first of these patients was suffering from dysphagia, and the other had EA type E diagnosed and operated on at age 4 years. He did not undergo preoperative echocardiography, and the ARSA was an incidental finding during the systematic review of esophagrams by radiologists. An angioscan and/or MRI was performed in 2 patients, confirming the presence and identifying the anatomy of the vascular malformation.

Most of the ARSAs were detected during the neonatal period before the onset of any symptoms. Three children (33%) with an ARSA remained completely asymptomatic during follow-up; the other 6 children experienced respiratory and/or digestive symptoms. **Table III** (available at www.jpeds.com) summarizes the clinical presentation of the 9 children with an ARSA and shows that in most cases, several confounding factors make it difficult to ascribe the symptoms to the ARSA. One child had a surgical correction of the ARSA and an aortopexy for severe tracheomalacia during the same surgery, making it difficult to interpret the reason for the resolution of respiratory symptoms after surgery. In 1 patient (patient 3), the role of the ARSA in dysphagia is likely, given that no esophageal stricture or esophagitis was demonstrated despite the presence of significant compression on the esophagram.

Factors Associated with Vascular Anomalies

A significant association between long-gap EA and vascular anomalies was found (5 of 14 [35%] vs 7 of 62 [11%]; OR, 4.37; 95% CI, 1.14-16.8; $P < .05$). Compared with children without a vascular malformation, those with a vascular malformation had a significantly higher proportion of severe cardiac malformations requiring surgery (5 of 14 [35%] vs 5 of 62 [8%]; OR, 6.33; 95% CI, 1.5-26; $P < .05$). Although the difference was not statistically significant, VACTERL was more frequent in children with vascular malformation (42% vs 21%).

Diagnostic Value of Upper GI Series

We retrospectively reviewed 254 upper GI series. The median number of esophagrams per patient was 2.5 (range, 1-14). According to our criteria, 154 of these upper GI series (60%) were conclusive for the specific detection of a vascular anomaly. A posterior indentation of the esophageal wall was present in 7 children, 6 of whom had a diagnosis of ARSA (n = 5) or ALSA (n = 1) demonstrated by echocardiography. Revision of the esophagrams revealed an ARSA in an asymptomatic child not known to have an aberrant vessel. In only 3 of these 7 patients, the indentation was previously mentioned in the medical imaging report. In 7 cases, the esophagram did not detect the vascular malformation. This finding was expected in 3 cases of RAA with mirror image, because the trajectory of the vessels is normal in this type of anomaly. The diagnosis of vascular malformation was missed in 4 cases (3 ARSA and 1 RAA with ALSA). Each of these 4 patients had a long-gap EA. In these 4 cases, the 12 upper GI series were inconclusive, as summarized in **Table IV** (available at www.jpeds.com). The sensitivity of the esophagram for the diagnosis of an ARSA or ALSA was 66%, the specificity was 98%, the negative predictive value was 95%, and the positive predictive value was 85%.

Discussion

We report that the overall incidence of ARSA and RAA is 18% in children who undergo surgery for EA/TEF. Long-gap EA and severe cardiac malformations requiring surgery are both significantly associated with vascular anomalies. We also show that the clinical consequences are variable, ranging from no symptoms to severe respiratory problems

and dysphagia. The diagnostic yield of routinely used techniques–preoperative cardiac ultrasound and esophagram–is not optimal.

Congenital vascular anomalies have been previously reported to be associated with EA/TEF (Table V)[4-9,11,12]; however, most previous studies focused on the presence of an RAA, which was considered a technical challenge during the initial repair of EA/TEF performed through a right-sided thoracic approach until recently.[4,7,8] In the present study, we found a 6% incidence of RAA in EA/TEF, in keeping with previous case series in which the incidence ranged between 1.8% and 13% (Table V).

Canty et al[13] reported an ARSA in 7 of 28 children with long-gap EA. Here we report a 12% incidence of ARSA in children with EA/TEF irrespective of the type of EA. This incidence is higher than that reported in the general population, which ranges between 0.5% and 1.8%.[14]

We found no cases of double aortic arch in the present series. This malformation appears to be rare, with only 3 cases of association of EA with a double aortic arch reported in the literature to date.[9,15,16]

An ARSA forms an incomplete vascular ring, but may lead to an extrinsic posterior esophageal compression. Indeed, because the ductus is generally on the left (same side as the aorta), the ring is not complete. Symptoms including dysphagia, recurrent aspiration, dyspnea, and cough have been reported. Dysphagia is the typical presenting symptom in adults[17,18]; respiratory symptoms can be seen in infants and children owing to the absence of tracheal rigidity.[19-22] Bakker et al[22] reported 15 children with an ARSA, including 6 without symptoms, 7 with dysphagia, and 10 with respiratory symptoms.

RAA is not considered a contraindication for the right-sided thoracic approach for primary repair in EA/TEF.[5,6] However, an RAA with an ALSA often forms a true and complete vascular ring (because the ductus generally remains on the left), and, in contrast to ARSA, this anomaly causes symptoms of dysphagia and respiratory distress in more than 50% of otherwise normal infants.[21,23,24]

Defining the liability of the vascular malformation is often impractical in the setting of children operated on for EA/TEF, because it may accompany various anomalies, including tracheomalacia, gastroesophageal reflux, anastomotic stricture, and congenital stenosis of the esophagus. Considering the possibility of an aortic anomaly in these patients is important because, far from being rare, these anomalies can be symptomatic, even dangerous. Lo et al[3] reported 2 cases of arterio-esophageal fistula formation and massive upper GI bleeding related to stent placement after EA repair, each with an unrecognized ARSA. Other authors have reported similar phenomena after prolonged (>17 days) use of a nasogastric tube in children without EA.[25] Situma et al[26] also demonstrated the importance of excluding a vascular anomaly before esophageal replacement surgery in children with EA/TEF.

Both long-gap EA and severe cardiac malformations necessitating surgery were significantly associated with vascular anomalies, confirming the findings of Canty et al,[13] who found an 43% incidence (12 of 28) of vascular malformation in children with long-gap EA vs a 2% incidence (2 of 175) in those with non–long-gap EA (Table V).

The association of aortic arch anomalies with severe cardiopathies and long-gap EA raises the hypothesis that a common mechanism may lead to these malformations. Neural crest defects, which are involved in digestive tract, aortic arch, and heart development,[27] may play a role, as has been suggested in an animal model.[28] Another suggested hypothesis is that the vascular malformation may impair esophageal development or worsen an actual EA.[13]

Unlike most previously published studies, here we report that echocardiography contributes to the diagnosis of ARSA but still may go unrecognized. The esophagram, performed routinely, is a valuable test for detecting an ARSA, as demonstrated by our sensitivity, specificity, and predictive value results. However, it is noteworthy that in some patients, the esophageal indentation was not described in the initial postoperative radiologic report, because the first esophagram was essentially dedicated to searching for anastomotic leaks, whereas subsequent ones focused on anastomotic strictures. Moreover, in most cases, the first examination did not include a true lateral position. Computed tomography and MRI are the most sensitive techniques for documenting great vessel anomalies and should be performed in patients with respiratory and/or digestive symptoms in whom the

Table V. Review of the literature on vascular malformations associated with EA

Authors	Year	Subjects with EA/TEF	Subjects with RAA	Subjects with ARSA	Diagnosis by echocardiography
Harrison et al[4]	1977	130	7	NS	1 of 6
Hartenberg et al[15]	1989	1	1	NS	0 of 1
Canty et al[13]	1997	28*	5	7	1 of 8
Bowkett et al[9]	1999	709	16	NS	1 of 7
Babu et al[8]	2000	476	12	NS	1 of 5
Allen et al[7]	2006	61	8	NS	5 of 8
Kanwal and Mondal[11]	2007	1	1	0	1 of 1
Bicakci et al[6]	2009	79	11	NS	1 of 10
Lo et al[3]	2012	2	0	2	0 of 2
Wood and Carachi[5]	2012	107	4	NS	1 of 4
Escobar et al[12]	2012	1	1	0	1 of 1
Berthet et al (present study)	2015	76	5	9	11 of 13

NS, not studied.
*Included only children with long-gap EA.

esophagram is inconclusive or when the condition indicates, to definitely rule out such malformations.

Surgical repair of a 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.[21,23,24] In adults with an ARSA, surgical vascular reconstruction is reserved for highly symptomatic patients possibly not responding to dietary modifications.[18] In children, the indication for surgical correction is less well established, but successful correction has been reported.[21,23,24]

The present study has some weaknesses. Because MRI and angioscan are the most sensitive methods available for detecting and finely describing the anatomy of aortic arch and vessel malformations, one can speculate on the possibility of missed diagnoses. Because all patients underwent EA/TEF repair and were studied with at least 1 esophagram, we are confident that no cases of RAA or complete vascular rings were missed. With regard to ARSA, it was actually not possible to definitively rule out the possibility of an ARSA, although we consider that clinical symptoms might not be attributable to an ARSA in absence of a posterior esophageal indentation on the esophagram.

In conclusion, vascular malformations are frequently associated with EA/TEF. ARSA and RAA have an incidence of 12% and 6%, respectively. Except in complete vascular rings, the liability of the vascular anomalies in respiratory and/or digestive symptoms may be difficult to demonstrate, because the symptoms may be attributable to tracheomalacia, gastroesophageal reflux, anastomotic stricture, or congenital stenosis of the esophagus. Echocardiography and the esophagram are complementary and effective tests, but their sensitivity is not optimal. Thus, using MRI or an angioscan to definitely rule out such malformations is recommended when stenting of the esophagus is indicated, before esophageal replacement surgery, and when prolonged (>2 weeks) use of a nasogastric tube is considered. ■

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Table II. Clinical presentation of patients with right aortic arch

Patient	Symptoms	Other conditions that can participate in the symptomatology	Surgical correction of vascular anomaly
1	Asymptomatic		No
2	Cyanosis with oxygen dependence Feeding difficulties	Cardiac anomalies resulting in death	No
3	Solid and liquid dysphagia Cough during feedings	Anastomotic stricture and congenital esophageal stenosis	Yes
4	Asymptomatic		No
5	Cyanosis with oxygen dependence Tracheostomy	Severe tracheomalacia TOF (operated) Anastomotic stricture (dilated)	Yes

TOF, tetralogy of Fallot.

Table III. Clinical presentation of children with ARSA

Patient	Symptoms	Other conditions contributing to symptomatology	Surgical correction of ARSA
6	Dyspnea Cyanosis with oxygen dependence Dysphagia	CoA Severe tracheomalacia Anastomotic and congenital esophageal stricture	No
7	Acute life-threatening events Respiratory infections	Severe tracheomalacia	Yes
8	Dysphagia without impaction		No
9	Oral aversion	Long gap Enteral feeding by gastrostomy	No
10	Asymptomatic		No
11	Asymptomatic		No
12	Severe dysphagia	Severe anastomotic stricture (with recurrence s/p multiple dilatations and surgery)	No
13	Dysphagia Cough with feedings	Tight anastomotic stricture No recurrence of fistula	No
14	Asymptomatic		No

CoA, coarctation of aorta.

Table IV. Representation of false-negative esophagrams and explanations

Patient	Type of malformation	Number of esophagrams	Number of conclusive examinations	Reasons
5	RAA + ALSA	2	0	Oblique image (n = 2) Nasogastric tube Significant postoperative anatomic alterations
6	ARSA	8	1	Oblique image (n = 6) Retrograde study via gastrostomy (n = 1) or nasogastric tube (n = 2) Leak image masking posterior wall of the esophagus (n = 3) Significant postoperative anatomic alterations
9	ARSA	3	1	Oblique image Leak image masking posterior wall of the esophagus Significant postoperative anatomic alterations
13	ARSA	2	1	Too little contrast Significant postoperative anatomic alterations (right-shifted mediastinum)

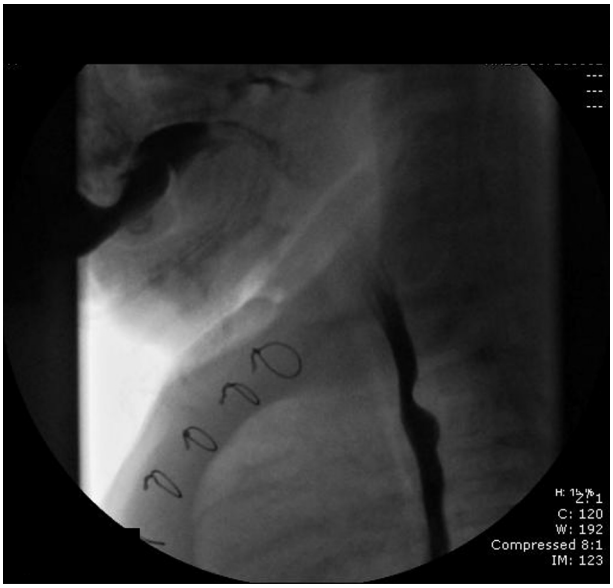

Figure 1. Posterior indentation of esophageal wall (patient 8, with ARSA).

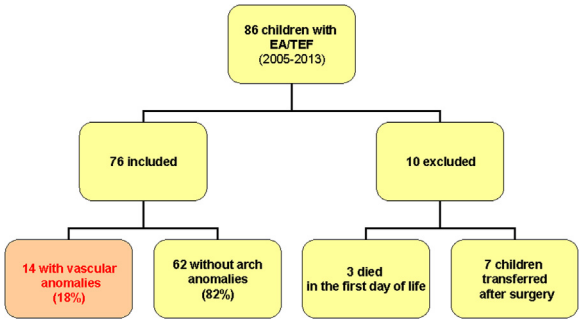

Figure 2. Flow chart of children with EA at Sainte Justine Hospital, January 2005 to October 2013.