CASE REPORT

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Esophageal Atresia Cases Concomitant with Persistent Left Superior Vena Cava Recognized Following Central Venous Catheter Insertion

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ABSTRACT Persistent left superior vena cava (PLSVC) is a rare but significant congenital vascular anomaly. It occurs when the left superior cardinal vein caudal to the innominate vein fails to regress. Its occurrence is usually isolated; however, studies have reported an increased frequency of related cardiac and extracardiac anomalies, particularly in connection with obstructive lesions of the left heart. Two cases of esophageal atresia, which have PLSVC anomaly recognized following insertion of a central venous catheter to the left internal jugular vein that seem to lie parallel to the aorta on the direct X-ray scan and proven by contrast-enhanced echocardiography later on, are presented. Esophageal atresia with PLSVC anomaly cases in English literature is summarized and differences in clinical presentations are discussed.

Keywords: Persistent left superior vena cava; esophageal atresia; central venous catheter

The first description of the persistent left-sided superior vena cava (PLSVC) was reported by Le Cat in 1738.[1] PLSVC is the most common abnormality of systemic venous return, with a prevalence of 0.1-0.5% in general population and up to 12.9% in patients with congenital heart disease. It is thought that this anomaly is seen more frequently in babies with esophageal atresia.[2] It is important to be aware of the presence PLSVC anomaly, especially when inserting a central venous catheter. We present 2 cases with esophageal atresia accompanying PLSVC anomaly recognized after catheter insertion.

CASE REPORT

CASE-1

A 1,420 g Syrian baby girl, born to 36 years old mother at 32 gestational weeks, delivered via cesarean section. The baby was admitted to the neonatal intensive care unit (NICU) due to prematurity and very low birth weight. The chest X-ray was obtained because the orogastric tube could not be inserted, and the findings were consistent with esophageal atresia (Figure 1a). Thoracoscopic repair for Type C esophageal atresia and laparotomy for duodenal atre-

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sia were performed on the 2nd postnatal day. On the postnatal 8th day, a 3F-60 mm central venous catheter was inserted from the left internal jugular under the guidance of ultrasonography. In the direct X-ray taken after the procedure, it was seen that the catheter was placed in the left paracardiac region (Figure 1b). The catheter was thought to be extravascular, but there was blood coming from the catheter. Echocardiography revealed persistent left superior vena cava anomaly that could not be detected in previous echocardiography. Due to the convenient location of the catheter, we decided to use it. No adverse events were reported related with the catheter. Unfortunately the patient died at 33 days of age due to septic shock. Informed consent was obtained from the parents.

CASE-2

The 2nd patient was a 1,555 g a baby boy, born to a 21 years old mother at 30 weeks gestation via cesarean section. The baby was admitted to the NICU due to prematurity and low birth weight. The chest X-ray was taken because the orogastric tube could not be inserted, and consistent findings with esophageal atresia were noted (Figure 2a). Thoracoscopic repair was performed for Type C esophageal atresia on the 2nd postnatal day. On the postnatal 9th day, a 3F-60 mm central venous catheter was inserted from the left internal jugular under the guidance of ultrasonography. In the direct X-ray taken after the procedure, it was seen that the catheter was placed in the left paracardiac region (Figure 2b). Given the previous experience with the very 1st case, it was thought that there might be a persistent left superior vena cava anomaly and echocardiography was performed immediately. PLSVC and coronary sinus dilatation were detected on echocardiography. The catheter was noted to pass from right atrium to the right ventricle so it was withdrawn to the appropriate place and was used uneventfully. The patient is still being followed in the pediatric surgery intensive care unit. Informed consent was obtained from the parents.

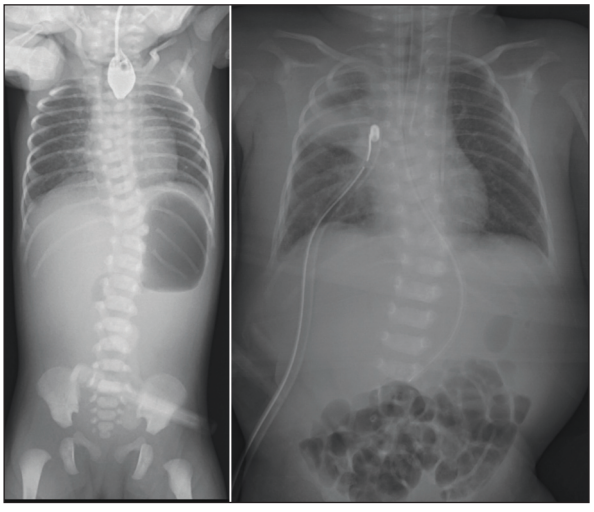

FIGURE 1: a) On the 1st day of the 1st case, direct X-ray taken with contrast in the esophagus. Esophageal atresia, hemi-vertebrae and double bubble appearance. **b)** Unusual position of the catheter tip aligning along with the aorta on radiograph

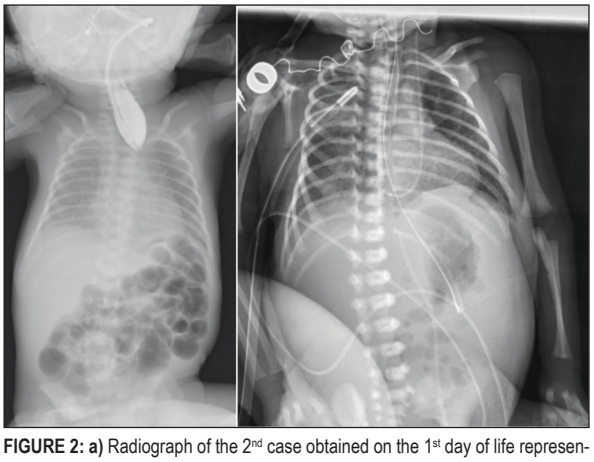

FIGURE 2: a) Radiograph of the 2nd case obtained on the 1st day of life representing esophageal atresia. **b)** Unusual position of the catheter tip aligning along with the aorta and finally placed in the right atrium on radiograph

DISCUSSION

The association of PLSVC with esophageal atresia is common. The incidence of vascular anomalies accompanying esophageal atresia is 9.9%.2 PLSVC usually drains into a dilated coronary sinus, but may occasionally drain into the left atrium.

Esophageal atresia is the most common congenital anomaly of the esophagus, occurring in 1 in 2,500-4,500 live births, and 50% of patients are accompanied by additional anomalies.[3] It usually occurs in the 4th week of pregnancy when there is a problem in the septation process of the trachea and esophagus. Many anomalies may accompany esophageal atresia. In a study, PLSVC was found in 8 of 89 esophageal atresia cases whose data could be accessed. No significant relationship was found be-

TABLE 1: An overview of the patients reported in the literature

	GW	BW	Gender	Atresia type	Other anomalies	Prognosis	Karyotype
1. Mowery, et al.[2]	-	-	-	1 case Proximal atresia distal atresia Others unknown	4/8 cases cardiac anomaly 5/8 cases renal anomaly 2/8 cases vertebral anomaly 3/8 cases skeletal anomaly 0/8 case gastrointestinal anomaly	-	-
9. Lai, et al.[4]	36+2	1,832 g	Female	Proximal atresia distal fistula	Pre-axial polydactyly	Died at postnatal 4 days	-
10. Esmer[5]	33		Male	-	Polyhydramnios, anal atresia, unilateral hypoplastic kidney	Surgery, survived, 6 years	Normal
11. Bergi, et al.[6]	Fetus	Fetus	Fetus	-	Mitral stenosis clinodactyly, strawberry sign, SUA, IUGR, polyhydramnios	Termination of pregnancy;	47,XY,+18
12. Arbell, et al.[7]	Term	3,000 g	Male	-	-	-	-
13. Avolio, et al.[8]	36	2,570 g	-	Proximal atresia distal fistula	None	-	-
14. Sajnach Menkea, et al.[9]	Fetus	Fetus	Fetus	-	PLSVC without bridging vein (bilateral SVC)	-	-
15. Broman, et al.[10]	Term	-	Male	-	None	Primary repair was performed in the neonatal period, and she was taken to ECMO as a result of hypoxemic respiratory failure secondary to sepsis when she was 5 months old and weighed 4.2 kg. Prognosis?	-
16. Nair et al.[11]	31+3	1,450 g	-	-	Imperforate anus, esophageal atresia, tracheal atresia	Exitus in the first hour of your life	-
17. Du et al.[12]	26	Fetus	Male	-	Lemon head, aplasia radii, ectrodactyly, clenched hands with overlapping fingers, SUA, cord cyst, polyhydramnios, VSD	-	47 XY+18
18. Galindo et al.[13]	28	-	-	Proximal atresia distal fistula	Truncus arteriosus	Esophageal and cardiac surgery was performed, died at the age of 5 months.	Normal
19. Case-1	32+2	1,420 g	Female	Proximal atresia distal fistula	Secundum ASD, Duodenal atresia, Hemivertebra	Died due to sepsis on postnatal 33rd day	-
20. Case-2	30+6	1,555 g	Male	Proximal atresia distal fistula	VSD	Still alive	-

GW: Gestational week (in weeks+days); BW: Birth weight; SUA: Single umbilical artery; IUGR: IntraUterin growth restriction; PLSVC: Persistent left superior vena cava; SVC: Superior vena cava; ECMO: ExtraCorporeal membrane oxygenation; VSD: Ventriculer septal defect; ASD: Atrial septal defect

tween other anomalies, genders and birth weeks-weights of these 8 cases.[2]

In our 2 cases, similar features were premature babies with esophageal atresia and PLSVC, as well as the diagnosis of PLSVC was proven by central venous catheter insertion in both cases. In both cases the presence of PLSVC was not noted in their 1st echocardiography obtained to screen additional structural hearth anomalies before the surgery performed. Recognition of PLSVC was possible only after the insertion of central catheter and the unusual position or the catheter on X-ray scan was noticed. There are 18 cases in the literature in which esophageal atresia and left superior vena cava coexistence have been reported. The characteristics of 20 cases in literature, together with our cases, are represented in Table 1.

Atrial septal defect, ventricular septal defect, atrioventricular septal defect, pulmonary stenosis or atresia, transposition of the great arteries, and conotruncal heart anomalies are congenital heart defects that may be associated with PLSVC. This association is explained by the failure of the left cardinal vein to regress adequately during embryological development.[1] In our patients, no associated cardiac anomaly was detected.

Some complications may occur after insertion of a catheter into the left superior vena cava. In a previous report in a preterm infant, a central venous catheter was found to be in the left superior vena cava using contrast-enhanced imaging methods. After a short time, the general condition of the patient deteriorated and he died.[14] None of the patients we reported had an adverse event related to catheter insertion in PLSVC.

Most of the babies with esophageal atresia need a central venous catheter because of long-term feeding problems. In a series reviewed, it was shown that more than 25% needed at least one central venous catheter during hospitalization.[15] It should be kept in mind that the frequency of persistent left superior vena cava in infants with esophageal atresia is more common than in the general population, therefore, preferably the right side should be used. However, the left superior vena cava can also be used for central venous access, but its complications should be closely monitored.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Mustafa Şenol Akın, Doğuş Çalışkan, Hayriye Gözde Kanmaz Kutman, Bekir Furkan Yalçın;* ***Design:*** *Mustafa Şenol Akın, Ahmet Vedat Kavurt, Hayriye Gözde Kanmaz Kutman, Bekir Furkan Yalçın;* ***Control/Supervision:*** *Mustafa Şenol Akın, Doğuş Çalışkan, Hayriye Gözde Kanmaz Kutman, Ahmet Vedat Kavurt, Bekir Furkan Yalçın;* ***Data Collection and/or Processing:*** *Mustafa Şenol Akın, Doğuş Çalışkan, Hayriye Gözde Kanmaz Kutman, Ahmet Vedat Kavurt, Bekir Furkan Yalçın;* ***Analysis and/or Interpretation:*** *Mustafa Şenol Akın, Hayriye Gözde Kanmaz Kutman, Bekir Furkan Yalçın;* ***Literature Review:*** *Mustafa Şenol Akın, Hayriye Gözde Kanmaz Kutman, Bekir Furkan Yalçın;* ***Writing the Article:*** *Mustafa Şenol Akın, Hayriye Gözde Kanmaz Kutman, Bekir Furkan Yalçın;* ***Critical Review:*** *Mustafa Şenol Akın, Hayriye Gözde Kanmaz Kutman, Bekir Furkan Yalçın;* ***References and Fundings:*** *Mustafa Şenol Akın, Hayriye Gözde Kanmaz Kutman, Bekir Furkan Yalçın;* ***Materials:*** *Mustafa Şenol Akın, Hayriye Gözde Kanmaz Kutman, Bekir Furkan Yalçın, Doğuş Çalışkan, Ahmet Vedat Kavurt.*

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