

Case Study

Title: "Imaging of congenital subclavian artery to subclavian vein fistula in a newborn"

Abstract:

The purpose of this paper is to report a congenital subclavian artery to subclavian vein fistula in a few weeksold male infant who presented with congestive heart failure and pulmonary hypertension. He had been scanned by various imaging modalities including echocardiography, cardiac Computed tomography (CT), diagnostic cardiac catheterization, cardiac magnetic resonance angiography (CMRA) and brainCT angiography. He had successful surgical ligation after failure of device occlusion trial at age of 9 months.

Keywords: congenital subclavian artery to subclavian vein fistula

Introduction:

An arteriovenous fistula (AVF) is an abnormal connection between an artery and a vein results in shunting of blood between the two, therefore bypassing the high-resistance capillary system which may be congenital or acquired. It was first recognized by Hunter in 1764. In general, AVFs are rare but correctable cause of hyperdynamic circulation and congestive heart failure as well as pulmonary hypertension when it develops in utero (1). Congenital systemic arteriovenous fistulae are generally rare anomalies in infants and children. AVFs in the head and neck region are rare. The majority of cases involving the major vessels of the neck are of traumatic origin, either blunt or penetrating. Other causes of these fistulas include ruptured aneurysms, collagen deficiency syndromes, arterial dissections, fibromuscular dysplasia, and extensive infection of the neck. Congenital arteriovenous communications involving the subclavian artery and jugular veins are rare (2, 3).

Acquired subclavian arteriovenous fistulae, which can result from trauma or iatrogenic causes, are rare because these vessels have good protection by the overlying bony skeleton, surrounding muscular structures and sheath (4). Iatrogenic subclavian arteriovenous fistula most commonly occurs after vascular access in catheterization procedures and can cause heart failure and be limb-threatening (4). Although many arteriovenous malformations (AVMs) are asymptomatic, they can

Comment [A01]: Please specify the age

Comment [A02]: computed

Comment [A03]: do not use abbreviations in the abstract

Comment [A04]: do not use abbreviations in the abstract

Comment [A05]: cranial

Comment [A06]: do not use abbreviations in the abstract

Comment [A07]: that results

Comment [A08]: please insert an appropriate reference

Comment [A09]: causes

Comment [A010]: also rarely encountered.

Comment [A011]: please insert an appropriate reference

Comment [A012]: You had abbreviated as AVF in the first paragraph, why do you still write as arteriovenous fistula???

33 cause severe pain, hemorrhage or can lead to other serious medical problems as pulmonary
34 embolism. In subclavian arteriovenous fistula, the initial symptoms and signs may be congestive
35 heart failure or continuous murmurs (4).
36

37 CASE REPORT:

38 A three weeks old male was referred to our hospital with the diagnosis of subclavian artery to
39 subclavian vein fistula. He was born with a spontaneous vaginal delivery prematurely at 34 weeks
40 gestation of single pregnancy with a birth weight of 1.8 kg. His APGAR score was 6 and 8 at 1 and 5
41 minutes respectively. He developed respiratory distress immediately after birth and diagnosed and
42 managed as neonatal respiratory distress syndrome treated with endotracheal surfactant, high
43 frequency oscillatory ventilation and received nitrous oxide for pulmonary hypertension (PHTN) for
44 some time. His family history was unremarkable and specifically negative for cardiopulmonary
45 disorders.
46

Comment [AÖ13]: The tense of the sentence has to be revised.

47 When admitted, he was mechanically ventilated; his vital signs showed oxygen saturation of 98% on
48 FiO₂ of 100% from the right arm; his blood pressure was 81/43 mmHg; and heart rate was 160 beats
49 per min. On physical exam he was found to have bounding peripheral pulses in both upper and lower
50 limbs, normal first and second heart sounds and a continuous machinery murmur, which was heard at
51 the left upper sternal border. Chest examination revealed normal vesicular breathing, equal on both
52 sides of the chest. The examination of abdomen, central nervous system, skin and mucosa did not
53 reveal any abnormality. Abdominal examination showed enlarged liver.
54

55 Chest roentgenogram demonstrated cardiomegaly. Electrocardiogram showed normal sinus rhythm,
56 normal QRS axis, and no right ventricular hypertrophy. Other laboratory findings showed
57 hemoglobin levels of 16 g/dl, normal liver function tests and renal profile with mildly elevated
58 ~~Prothrombin Time (PT)~~, Partial Thromboplastin (PTT) and International Normalized Ratio (INR).

Comment [AÖ14]: Prothrombin time and INR are the same things !!

59 An echocardiographic study showed situs solitus, levocardia, D-looped ventricles, and normally
60 related great arteries. It also showed a dilated right side of the heart with moderate tricuspid valve
61 regurgitation with peak systolic pressure gradient (PSPG) of 75 mmHg. There were 2 mm
62 arteriovenous fistula between the subclavian artery and vein with PSPG of 50 mmHg with dilated
63 superior vena cava as well as subclavian vein and artery with continuous flow in the subclavian
64 artery and diastolic run off in the descending aorta (Figure 1 & 2 Video 1).

Comment [AÖ15]: AVF

65

66 To further outline the lesion, cardiac CT was done the next day of admission and showed
67 arteriovenous fistula connecting the left subclavian artery to the large subclavian vein at the level of
68 the middle half of the clavicle (Figures 3-6). Cranial ultrasound was normal initially with no
69 hemorrhage or hydrocephalus.

70 The patient underwent cardiac catheterization with an attempt to coil occlude the fistula. The cardiac
71 catheterization confirmed the presence of AVM (Videos 2A& B). However due to the close
72 proximity of the subclavian artery and vein, presence of vital branches and the concern that any type
73 of device would occlude some of these vital branches and can cause progressive intra vascular
74 thrombosis; the intervention was not performed and the case was deferred to the surgical team. Due
75 to his small size, the operation was postponed and the child was followed conservatively with anti-
76 failure measures and will need ligation at age 7-8 months if clinical condition permits. Cardiac
77 magnetic resonance angiography (CMRA) was done at the age of 7 months and showed
78 evidence of left subclavian artery to the left subclavian vein fistula identified (Figure 7). Because the
79 infant had delayed neurological development; brain CT angiography was done at the same time and
80 showed abnormally dilated venous varix in the medial aspect of the middle cranial fossa (Figure 8).

81 Surgical ligation of the fistula was done at age of 9 months with excellent results with complete
82 anatomic correction and rapid resolution of right sided cardiomegaly and pulmonary hypertension.
83 The dilated intracranial venous system (which resulted likely from chronic venous hypertension) was
84 significantly improved following fistula ligation. He continued however to have brain atrophy with
85 extra axial fluid collection.

86

87 Discussion:

88

89 An arteriovenous fistula (AVF) is an abnormal direct vascular channel between an artery and a vein
90 that results in shunting of blood between the two without an intervening capillary network (2). AVF
91 is either congenital or acquired. The congenital type is frequently more anatomically complex and is
92 often too widespread for device occlusion and often need surgical intervention. Congenital fistulas
93 are composed of the embryonic mesoderm that result from an arrest in the embryological
94 differentiation during the development process of the vascular system with failure of the primitive
95 plexiform structures to differentiate normally into mature capillary and venous networks. This leads
96 to presence of abnormal arteriovenous channels; which could be single or multiple, local or diffuse,

Comment [A016]: In order to

Comment [A017]: performed

Comment [A018]: AVF

Comment [A019]: AVF not AVM

Comment [A020]: referred

Comment [A021]: what do you mean?

Comment [A022]: How did you decide? Is there any reference for that?

Comment [A023]: performed

Comment [A024]: You abbreviated it before!!

Comment [A025]: You used the same sentence in introduction, do not repeat it here.

97 and visceral or cutaneous (5). It was also noted that there is higher incidence of AVF involving right
98 subclavian artery more than the left which could be related to the complexity of the embryologic
99 development of the right subclavian artery (6).

100

101 The volume and velocity of blood flow shunted through the fistula will depend on size of a fistula
102 and its proximity to the heart and the pressure gradient. The near to the heart and the wider is the
103 fistula, the more the increase in cardiac output and the more will be the cardiac work. For example, a
104 small fistula from a branch of the aortic arch may result in more cardiac work than a larger one in the
105 leg or forearm (7). Despite congenital AVF are rare but they are important causes of heart failure in
106 newborns, infants and young children. In our case the infant presented with high cardiac output heart
107 failure which denoted significant shunting of blood from the high pressure subclavian artery to the
108 low pressure subclavian vein through AVF.

109

110 There are few reports about AVF involving the subclavian artery. The fistula may connect the
111 subclavian artery with subclavian vein, innominate veins or Internal Jugular vein (8, 9). These
112 fistulae have a wide range of clinical manifestations from being asymptomatic to present with frank
113 heart failure. Local examination may reveal a continuous bruit in the upper chest, loudest at the right
114 or left infra-clavicular area (10). Congenital AV fistula usually present with heart failure in newborn,
115 infants and young children. Pulmonary hypertension can result from giant AFV (11). However,
116 congenital AV fistula involving subclavian artery and vein was previously reported in 2 adults (12,
117 13).

118

119 There are different diagnostic modalities that help in diagnosing these types of fistulae. X-ray may
120 show variable degrees of cardiomegaly with possible increased pulmonary vascular markings,
121 widened mediastinum and prominence of the superior vena caval shadow. The cardiac silhouette may
122 be similar to the "snowman" or "hourglass" configuration seen in older patients with total anomalous
123 pulmonary venous return. However, the uncommon location of the fistula may produce other
124 radiologic findings and can be misleading. ECG may be normal or show right axis deviation, right
125 and left atrial enlargement and right ventricular hypertrophy pattern. Sometimes ECG shows left
126 ventricular hypertrophy (9, 13).

127

128 Two-dimensional echocardiography with color Doppler imaging may aid the initial diagnosis.
129 Various echocardiographic modalities can help in evaluation of AVF. M-mode may show dilated

Comment [A026]: AVF

Comment [A027]: Please insert an appropriate reference.

130 right atrium, right ventricle, left atrium, and/or left ventricle. Adequate echocardiographic
131 examination can efficiently visualize the fistula however, in some cases it may be difficult and other
132 diagnostic tools may be needed; at least to confirm the diagnosis (14).

133

134 Computed tomographic and magnetic resonance angiography can provide images of the anatomy of
135 the arteriovenous communication, typically with early contrast filling in the vein during the arterial
136 phase. Detailed anatomical views of the involved vessels and the site and size of the AVF is helpful
137 when making decisions about the best treatment option. Cardiac CT scan has increased risk of
138 ionizing radiation but it has very short study time, its spatial resolution is as little as 0.4 mm.
139 Meanwhile, the cardiac magnetic resonance imaging (CMRI) can acquire cine image in any desired
140 plane without the risk of ionizing radiation; available post-processing tools and real time images can
141 be obtained without intravenous contrast. However, CMRI has long study time and its long axis
142 resolution is just below 1 mm (15). Time-resolved non-contrast enhanced 4-D dynamic magnetic
143 resonance angiography can provide detailed information on the location and size of the fistula and
144 the dynamic flow of blood similar to what we can get by invasive angiograph. Angiography during
145 cardiac catheterization is not usually required for routine diagnosis but may be used at the time of
146 therapeutic procedures or when other non-invasive imaging fails to identify this abnormality (13).

147

148 Closure of the fistula is recommended because of their tendency to become hemodynamically more
149 significant with time, the risk of bacterial infection, pulmonary embolism and thrombosis and the
150 possibility of developing aneurismal or degenerative changes or even rupture. If amenable, trans-
151 catheter occlusion of the fistula can be done, however, subclavian fistula is difficult for trans-catheter
152 occlusion because of the close proximity to the heart and the major vessels. Nevertheless, surgery is
153 usually the recommended treatment for either symptomatic or non-symptomatic arteriovenous
154 fistulas if the fistula is not amenable to trans-catheter occlusion or after its failure as in our case. The
155 choice of treatment is determined by the location and anatomy of the lesion (13, 16).

156

157 **Conclusion:**

158 Congenital systemic arteriovenous fistulae are generally rare anomalies in infants and children
159 including that in the head and neck region. In subclavian arteriovenous fistula, the initial symptoms
160 and signs may be congestive heart failure or continuous or innocent murmurs and can present very
161 early during the neonatal region or late during adulthood. There are different diagnostic modalities
162 including Two-dimensional echocardiography with color Doppler imaging, computed tomographic

Comment [A028]: So you have to convince me why you waited until 9 months of age !!!!

163 and magnetic resonance angiography and conventional angiography. Closure of the fistula is
164 indicated to avoid many complications including heart failure, infection or aneurismal dilations.

165

166 **COMMENTS**

167 **Case characteristics**

168

169 Three weeks old male was referred to our hospital with the diagnosis of subclavian artery to
170 subclavian vein fistula. Documented clinical presentation and progress, imaging findings and
171 operative treatment were investigated.

172

173 **Clinical diagnosis**

174 The baby presented with respiratory distress immediately after birth and diagnosed and managed as
175 neonatal respiratory distress syndrome. After, that signs of heart failure with bounding peripheral
176 pulses in both upper and lower limbs, normal and a continuous machinery murmur, which were heard
177 at the left upper sternal border.

178

179 **Imaging diagnosis**

180

181 Chest roentgenogram demonstrated cardiomegaly. Echocardiographic study showed a dilated right
182 side of the heart, moderate tricuspid valve regurgitation and 2 mm arteriovenous fistula between the
183 subclavian artery and vein with PSPG of 50 mmHg with dilated superior vena cava as well as
184 subclavian vein and artery with continuous flow in the subclavian artery and diastolic run off in the
185 descending aorta. Cardiac magnetic resonance angiography (CMRA), Cardiac CT and cardiac
186 catheterization was done and confirms the diagnosis.

187

188 **Differential diagnosis**

189

190 Subclavian artery to subclavian vein fistula should be differentiated from causes of hyperdynamic
191 heart failure, other congenital arteriovenous fistulas and total anomalous pulmonary venous drainage.

192

193 **Treatment**

194

195 Trans-catheter occlusion if amenable, surgical closure will be determined by the location and
196 anatomy of the lesion

197 **Related reports**

198

There are few reports about AVF involving the subclavian artery.

Experiences and lessons

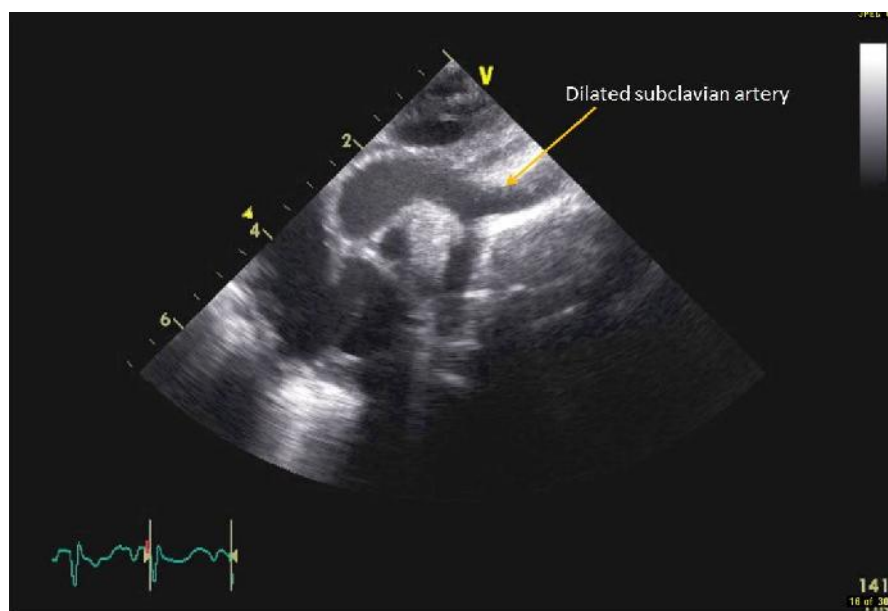
Closure of the fistula is indicated to avoid many complications including heart failure, infection or aneurismal dilations.

References:

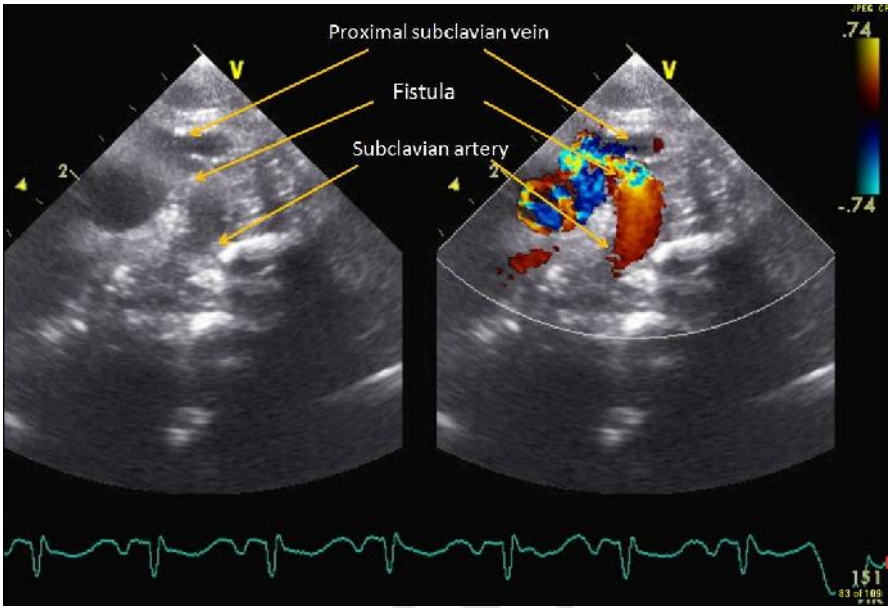
1. Burry M, Reig AS, Beierle EA, Chen MK, Mericle RA. Extracorporeal membrane oxygenation combined with endovascular embolization for management of neonatal high-output cardiac failure secondary to intracranial arteriovenous fistula. Case report. *J Neurosurg.* 2004 Feb;100(2 Suppl Pediatrics):197-200. DOI: 10.3171/ped.2004.100.2.0197. PMID: 14758950
2. Chi M, Wang N, L Yu. Treatment of a Rare Congenital External Carotid Arteriovenous Fistula with Transcatheter Coil Embolization. *ActaCardiol Sin* 2010;26:272_5Acta Cardiol Sin. 2010;26:272_5.
3. Anastacio MM, Castillo-Sang M, Smith TW, Saad N, Lawton JS. Iatrogenic left internal thoracic artery to left subclavian vein fistula after excimer laser pacemaker lead extraction. *J ThoracCardiovasc Surg.* 2012 Apr;143(4):e35-7. doi: 10.1016/j.jtcvs.2012.01.014. PMID: 22306221
4. Ding M, Hu J, Ni J, Lv H, Song D, Shu C. Iatrogenic subclavian arteriovenous fistula: rare complication of plate osteosynthesis of clavicle fracture. *Orthopedics.* 2012 Feb 17;35(2):e287-9. doi: 10.3928/01477447-20120123-21. PMID: 22310422
5. Duncan JM, Cooley DA, Vargo TA, Gutgesell HP. SUCCESSFUL SURGICAL MANAGEMENT OF CONGENITAL ARTERIOVENOUS FISTULA OF THE SUBCLAVIAN ARTERY: CASE REPORT. *Cardiovasc Dis.* 1978 June; 5(2): 139-143. PMID: 15216065
6. Stanley AC, Menzoian JO. A Congenital Arteriovenous Malformation of the Right Subclavian Artery. *VASC ENDOVASCULAR SURG.* 2000; May/June;34 (3) 271-5. doi: 10.1177/153857440003400315
7. Pagel A, Bass A, Strauss S, Peleg E, Rapoport MJ. High output cardiac failure due to iatrogenic A-V fistula in scar: a report of a case and review of the literature. *Cardiovasc Surg.* 2003 Aug;11(4):317-9.
8. Dogan R, Yilmaz M, Ozkutlu S, Elsharshari H. Congenital subclavian artery to subclavian vein fistula presenting with congestive heart failure in an infant. *PediatrCardiol.* 2000 May-Jun;21(3):269-70. DOI 10.1007/s002460010056. PMID: 10818190
9. Sapire DW, Lobe TE, Swischuk LE, Casta A, Schwartz MZ, Droge M. Subclavian-artery-to-innominate-vein fistula presenting with congestive failure in a newborn infant. *PediatrCardiol.* 1983 Apr-Jun;4(2):155-7. PMID:6348716
10. Balakrishnan PL, Guleserian KJ, Schuster J, Blalock SE. Congenital arteriovenous malformation between the subclavian artery and superior vena cava presenting in neonatal heart failure. *PediatrCardiol.* 2011 Dec;32(8):1204-6. doi: 10.1007/s00246-011-9990-z. PMID:21509497.
11. Muñoz IA, Bustos GA, Pardo AG, García EM, Del Mar Molinero-Casares M, Ruiz-Canela JJ, Ramírez VM. Heart failure and severe pulmonary hypertension secondary to a giant renal arteriovenous malformation. *J Ultrasound Med.* 2006 Jul;25(7):933-7.
12. Wong CK, Lau CP, Leung WH. An arteriovenous fistula from an aberrant right subclavian artery to the superior caval vein in a congenitally myxoedematous adult. *Int J Cardiol.* 1989; Oct. 25 (1):126-9.
13. Anoop TM, Sreejith P, Thomas JK, Gailin B, Jabbar PK, Ittycheria CC, George R. Congenital Subclavian Arteriovenous Malformation Causing Cardiac Failure in an Adult. *J Am Soc Echocardiogr.* 2009 Jul;22(7):864.e5-6. doi: 10.1016/j.echo.2009.03.011. PMID: 19398189
14. Tatum GH, Newman B, Ralphe JC. Neonatal congestive heart failure due to a subclavian artery to subclavian vein fistula diagnosed by noninvasive procedures. *Congenit Heart Dis.* 2006 May;1(3):120-3. doi: 10.1111/j.1747-0803.2006.00019.x. PMID: 18377556
15. Crean A. Cardiovascular MR and CT in congenital heart disease. *Heart.* 2007 Dec;93(12):1637-47. . doi: 10.1136/hrt.2006.104729. PMID: 18003697

16. Lookstein RA, GullerJ.. Embolization of complex vascular lesions. *Mt Sinai J Med*. 2004 Jan;71(1):17-28. PMID: 14770247

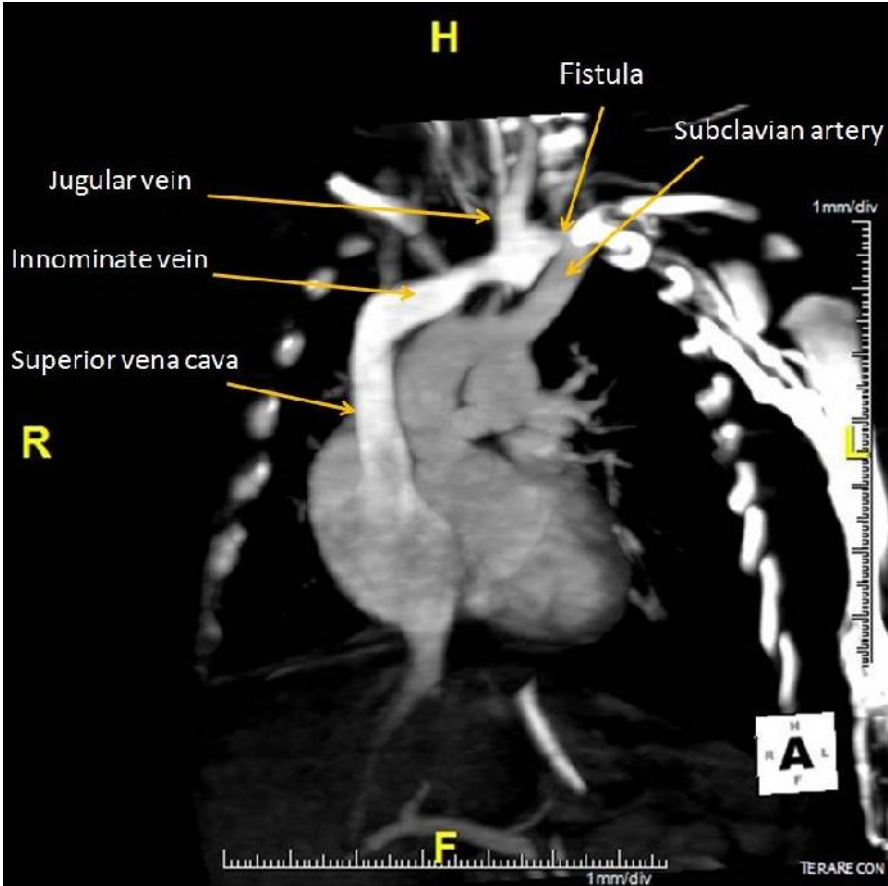
Figure 1: 2-D Echocardiogram showing the aortic arch with dilation of the left subclavian artery.



262 Figure 2: 2-D Echocardiogram with color flow Doppler showing dilated left subclavian artery
263 with the fistula connecting the left subclavian artery and vein.

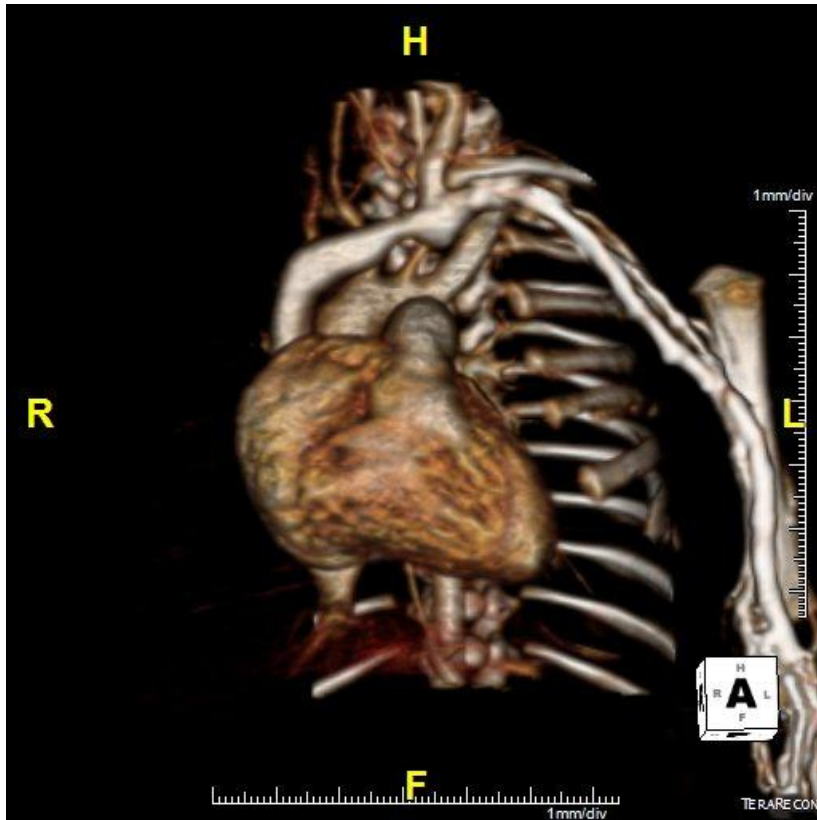


276 Figure 3: CTA image showing the site and size of the fistula with dilated innominate vein, left
277 subclavian artery, jugular vein and superior vena cava.



286 Figure 4: 3-D CTA reconstruction showing dilated innominate vein, left subclavian artery,
287 jugular vein and superior vena cava and the relation to the clavicle. You can also see the dilation
288 of the venous system of the left arm.

289



290

291

292

293

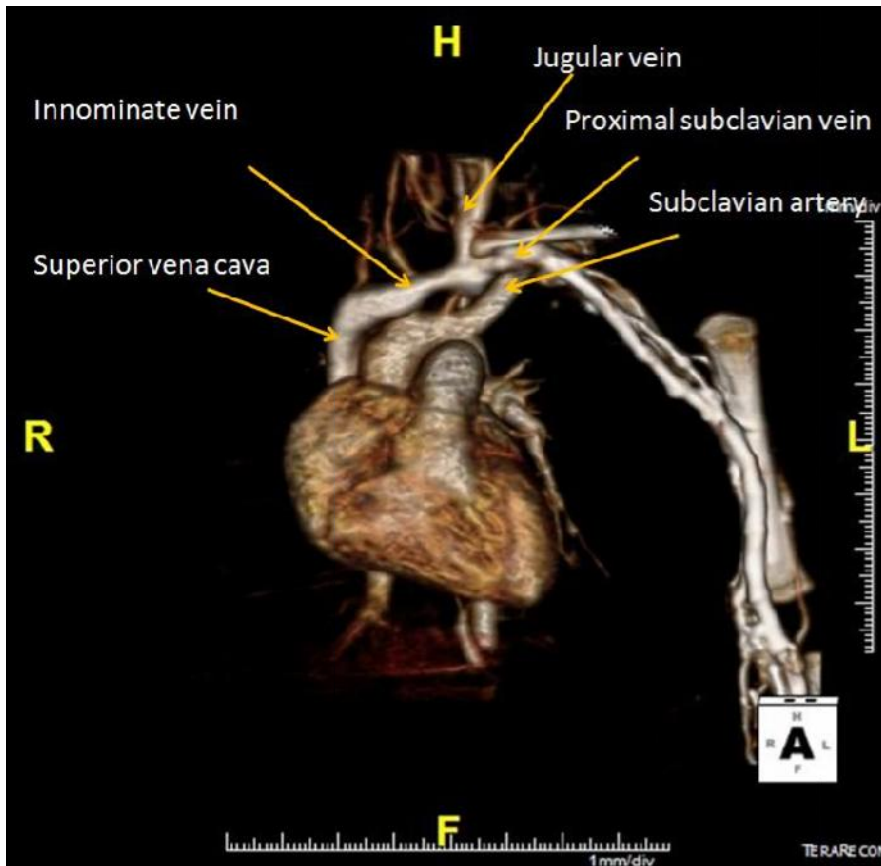
294

295

296

297

298 Figure 5: 3-D CTA reconstruction showing dilated innominate vein, left subclavian artery,
299 jugular vein and superior vena cava and the relation to the clavicle. You can also see the dilation
300 of the venous system of the left arm.



301

302

303

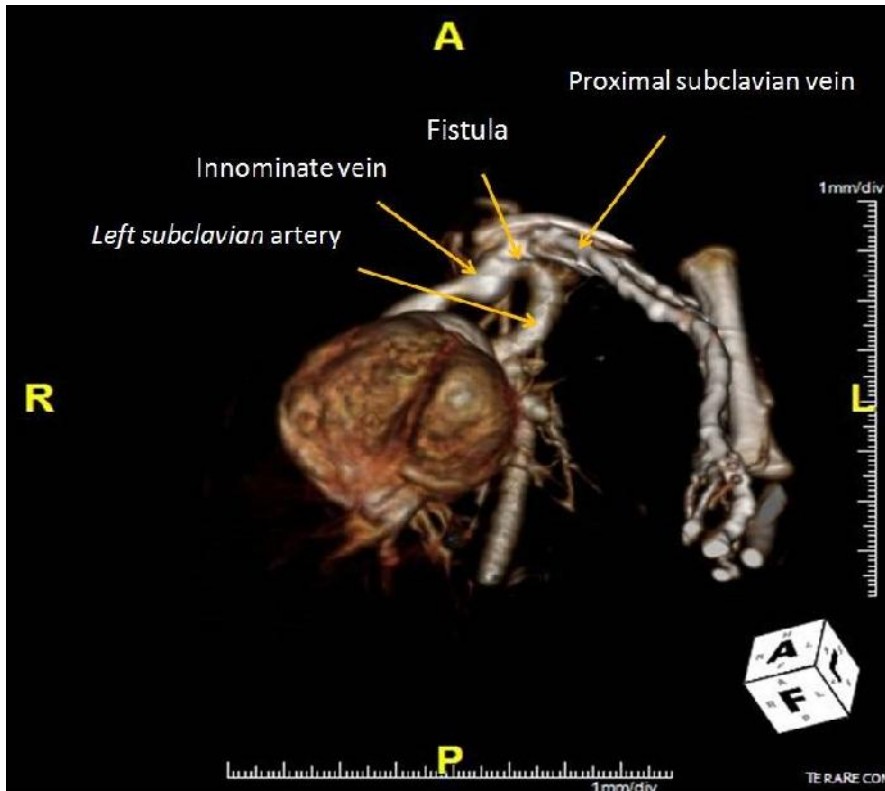
304

305

306

307

308 Figure 6: 3-D CTA reconstruction3-D CTA reconstruction showing the site of the fistula.



309

310

311

312

313

314

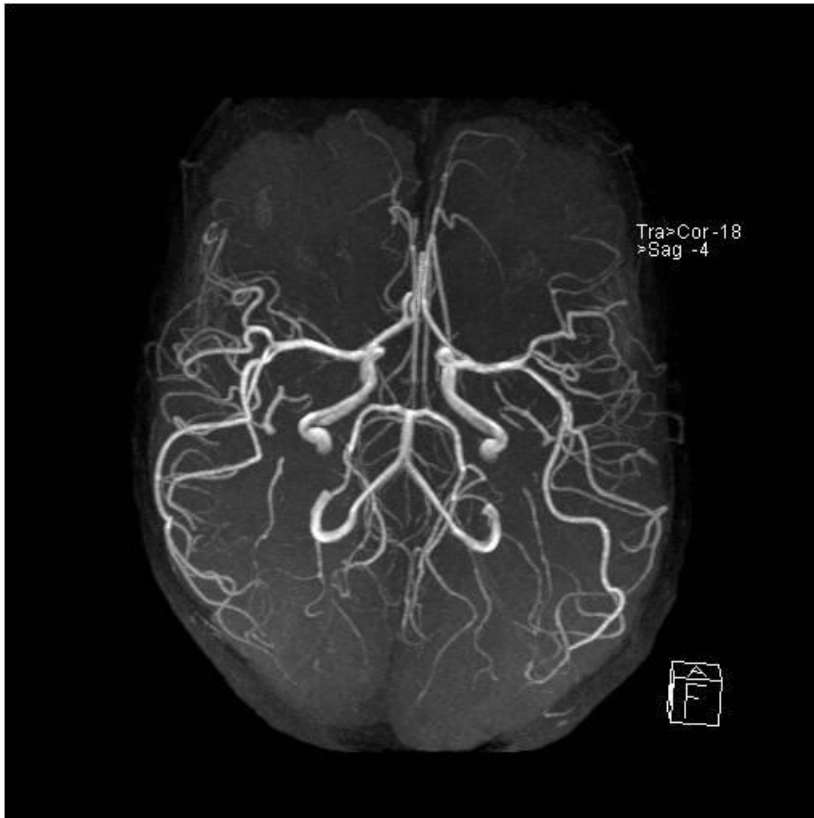
315

316

317

318

319 Figure 7: MRA neck vessels showing dilated left subclavian artery, internal jugular vein with
320 dilated intracranial veno-venous collaterals (the innominate vein did not fill possibly due to
321 thrombosis following central line placement).



322
323
324
325
326
327
328
329

330 Figure 8: CTA brain showing no other abnormal arterio-venous connections. In this arterial
331 phase you do not appreciate the venous system dilation or plexus of network.

332



333

334

335

336 **Video 1:** 2-D Echocardiogram with color flow Doppler of Suprasternal view showed dilated left
337 subclavian artery with the fistula connecting the left subclavian artery and vein.

338

339

340 **Video 2** A & B: Left ventricular angiogram demonstrating fistula between left subclavian artery
341 and vein. Note rapid filling of superior vena cava and right atrium. The lateral view showed
342 clearly the fistulous tract between left subclavian artery and vein.

343



Video 1 Fistula.avi

344

345

346



Video 2 A.avi

347

348



Video 2 B.avi

349

350

UNDER PEER REVIEW