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A Severe and Fatal Type A Aortic Dissection in an Adult with a Repaired Tetralogy of Fallot

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Abstract: We report a case of a 44-year-old woman surgically treated for tetralogy of Fallot who experienced an acute and extensive Stanford A type aortic dissection despite the meticulous follow-up. While aortic dilatation is prevalent in individuals with repaired tetralogy of Fallot, aortic dissection represents a rare consequence, that when it appears, is progressive and usually detected during the check-up visits. In the case reported, the dissection was unexpected and severe, and the patient's clinical state worsened suddenly, leading to death after a few days. Constant awareness for aortic aneurysms is essential in the Fallot tetralogy population, nevertheless, several causes may contribute to the acute worsening of the clinical condition until the patient's death.

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Introduction

Tetralogy of Fallot (TOF) is one of the most common cyanotic congenital heart diseases, that can be corrected by surgery: in fact, a radical correction is most often performed within one year of life.

In the scientific literature, there is an increase in cases of aortic involvement in many congenital heart diseases, especially in patients later after the repair of TOF such as dilatation and aortic dissection; patients with TOF have more residual findings in adulthood: pulmonary regurgitation and aortic root dilatation which may require surgical management in adulthood. Dilatation of the aorta is also frequent in TOF (up to 69%), however aortic aneurysm (with diameter ≥ 50 mm) was found in 9% only and aortic dissection is very rare in TOF (0–0.06%).

Different post-operative problems can occur and require some extra treatments, or a strict follow-up is always required (Vaikunth et al., 2022). Given the residual findings in the patient of this case, she requires lifelong follow-up by a cardiologist (Grotenhuis et al., 2018).

This case report describes the imaging examination, especially echocardiogram and computed tomography (CT) scans, in a patient who had an acute and severe aortic dissection, with an important hypotensive shock (Kim et al., 2011; Chow et al., 2020).

Case report

A 44-year-old patient came to the Emergency Department transported by ambulance in an unconscious state. The patient's condition immediately appeared severe: the skin was pale, sweaty, and cold; the pulse was extremely weak and there was arterial hypotension.

Given the critical condition, a consultation with the cardiologist was required and a transthoracic echocardiogram was performed. The latter demonstrated an

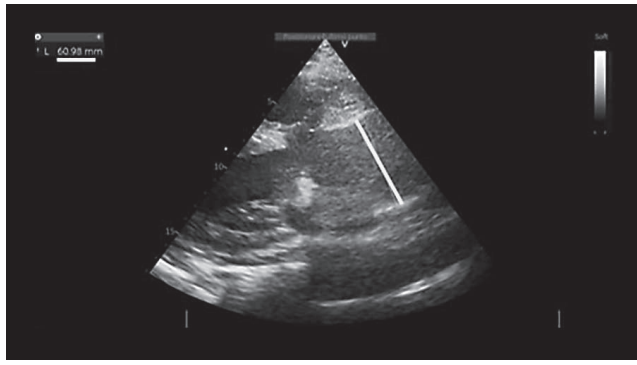

Figure 1 – The transthoracic echocardiography showed the aortic aneurysm with a severe dilatation of the aortic root (about 61 mm).

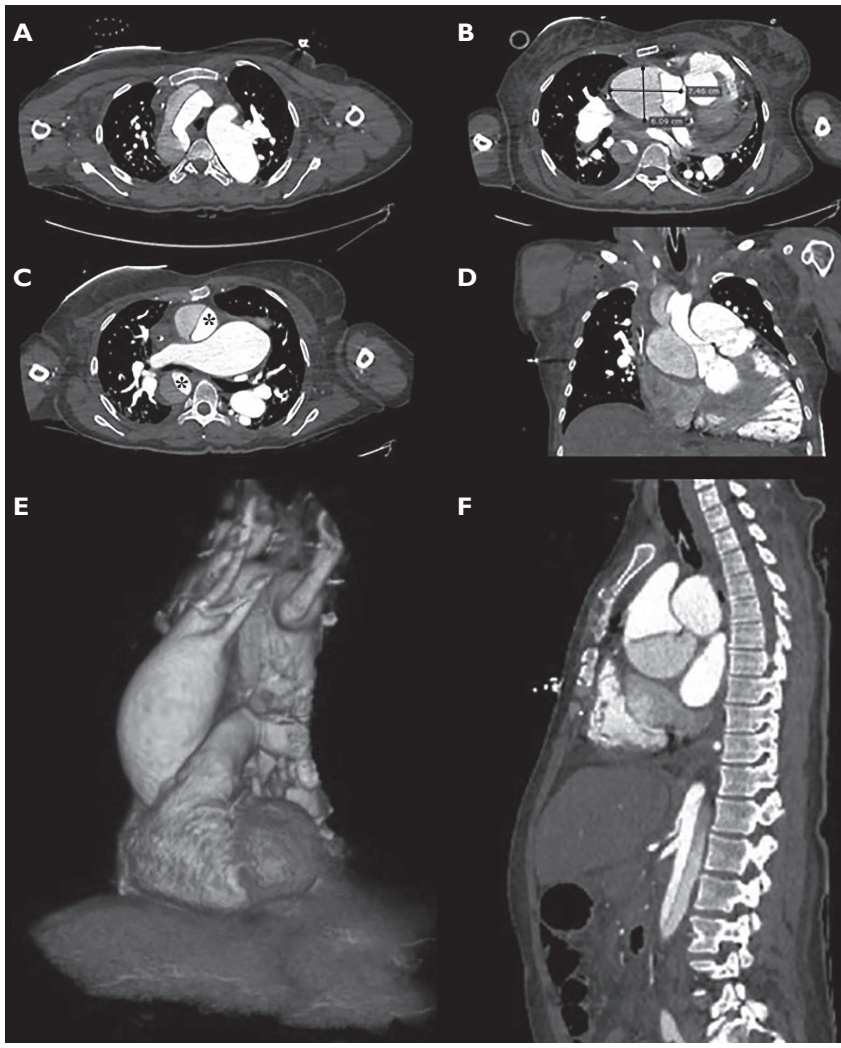

Figure 2 – Computed tomography angiography scan revealed a marked aneurysm of the aorta (A) (maximal dimension of 6×7 cm at the ascending aorta – B), with an intimal flap of dissection involving the aortic root, the aortic arch (C, D, E, F), and the right-sided descending aorta and dividing the true lumen (TL) – the star (C), from the false lumen (FL). There is a severe dilatation also of the ascending aorta (arch and descending are involved too). This dilatation is due to the dissection and the formation of a second lumen (false lumen) from the valvular plane and aortic root.

aneurysmal dilatation with a maximal diameter at the aortic root, resulting in severe aortic insufficiency (Figure 1).

The examination also revealed an intimal flap suspicious of aortic dissection and the presence of a pericardial effusion. After stabilization manoeuvres, the patient was immediately taken to the Radiology Department where a computed tomography

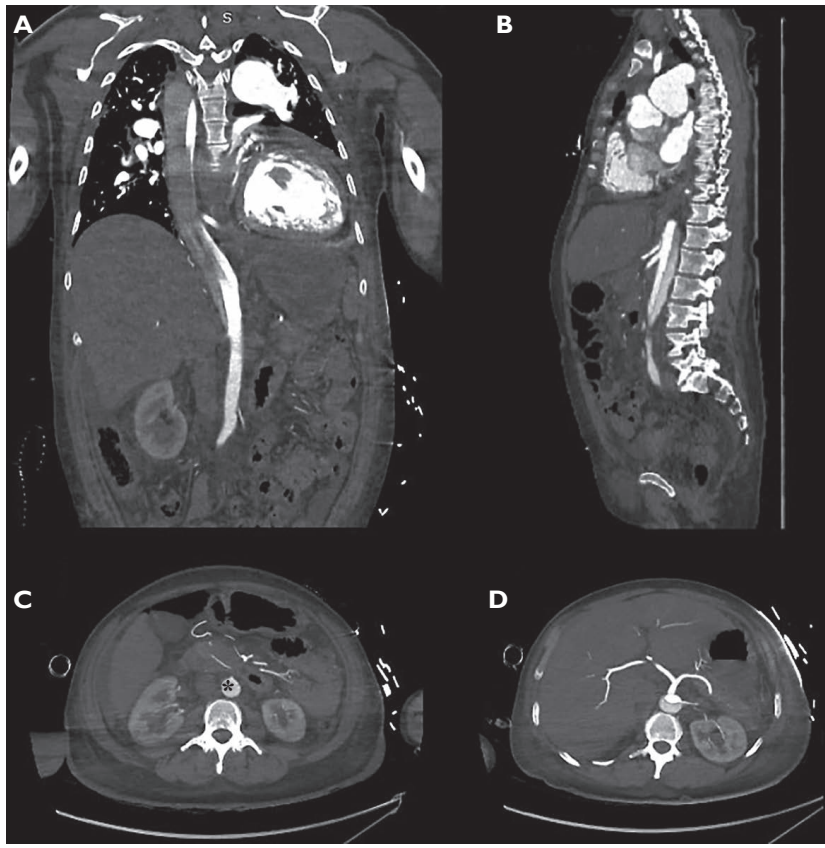

Figure 3 – Computed tomography angiography scan images, showing in coronal (A) and sagittal (B) the extension of the dissection flap extended to the upper limits of the iliac carrefour (shown in axial section, the lower limit of abdominal aortic dissection – C). The coronal section of the artery (B) is not entirely viewable given the scoliotic course of the abdominal aorta. With increased maximum intensity projection, the integrity of the celiac trunk is shown (D).

angiography (CTA) examination was performed. The patient had not been under cardiological control for many years and the aortic size was unknown before the dissection. The CT scan before and after contrast agent administration showed a severe aortic dissection, Standford type A, originating from the aortic root, with a dissection flap detected near the aortic valve, and reaching the abdominal aorta until its bifurcation in the common iliac arteries (Figures 2 and 3).

The Bentall operation was required in case of complicated aortic dissection, severe clinical condition, and CT findings of a type A dissection was not soon performed as the patient was inoperable at that moment and the surgery was delayed due to rather critical clinical conditions: visceral mal-perfusion and deterioration of the neurological state (Zhang et al., 2022; Shehata et al., 2023).

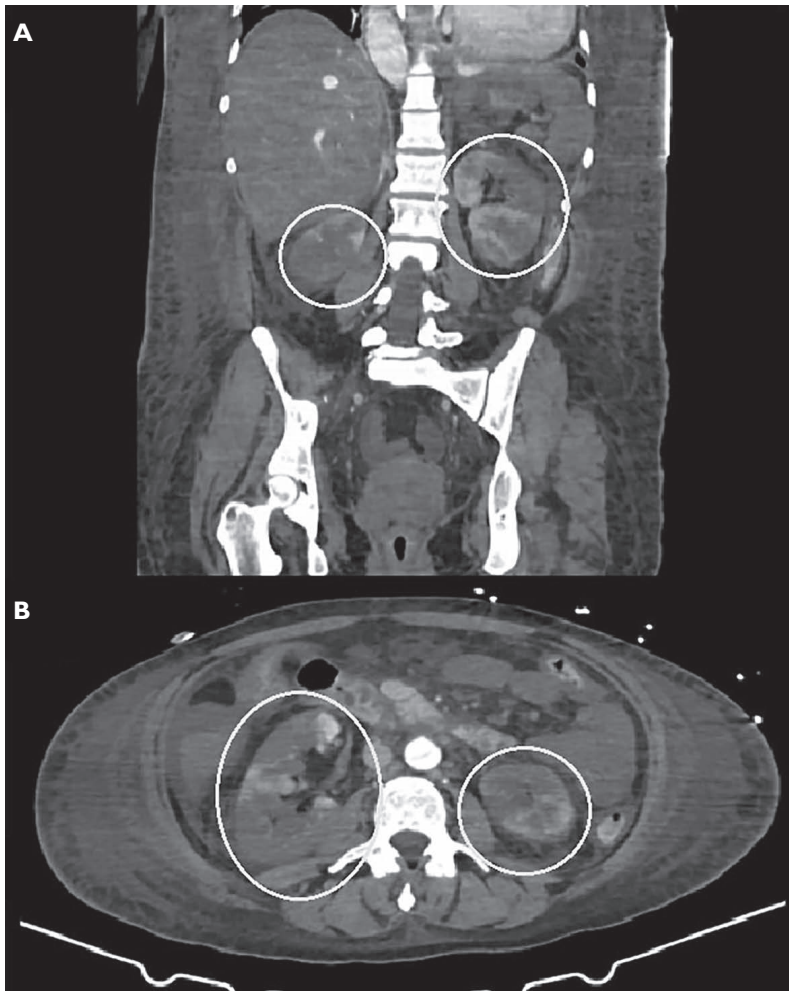

Figure 4 – Computed tomography angiography scans in coronal (A) and axial (B) planes, showed a severe condition of hypotension, with the contrast medium in the aortic vascular lumen even in delayed scans and abdominal visceral mal perfusion, especially liver and kidneys (circled the kidneys, in an end-stage condition due to hypotension), associated with a massive free abdominal fluid and oedema of soft tissues.

Therefore, the patient was hospitalized trying to stabilize volemia and clinical parameters; devices of central venous catheter (CVC), endotracheal tube (ETT), and a nasogastric feeding tube (NG) were implanted; and medical treatment was administered. After two days the CT scans were performed and confirmed the mal perfusion syndrome due to the severe dissection (Figure 4).

Due to the worsening of the clinical conditions of the patient, the surgery was never performed since she died in the days that followed.

Discussion

One of the most frequent congenital cardiac abnormalities is TOF consisting of the coexistence of pulmonary artery stenosis, the subaortic ventricular septal defect, and an anterosuperior deviation of the aortic root and sub-valvular right ventricular hypertrophy (Niwa, 2005; Kim et al., 2011; Vaikunth et al., 2022).

Distension of the aortic root, more commonly, and aortic dissection, more rarely, have been identified as complications following TOF repair, one of the most successful surgical corrections in treating congenital cardiac disorders. According to the literature, the risk factors for aortic dissection in TOF were age over 60 years, men's gender, hypertension, and of course the aortic diameter. In this case, there was a female and young patient, with an anamnesis not totally clear (except for corrective surgery done within the first year of life, as reported by relatives).

Aortic root aneurysm in TOF is a long-term clinical issue since a significantly dilated aorta can produce aortic regurgitation, dissection, or rupture, all of which can be fatal and necessitate surgical intervention (Zhang et al., 2022; Shehata et al., 2023).

The probable cause of dilatation is a congenital anterosuperior deviation of the aortic root with seeding over the ventricular septal defect however, the real mechanism of aortic root dilation remains unknown (Seki et al., 2016; Egbe et al., 2019).

Aortic dissection is a rarer entity in individuals with repaired TOF and when occurs it appears progressive in time, allowing a rapid detection and prompt medical treatment during the follow-up. The role of imaging, and CTA is essential in the evaluation of the aortic condition.

In the case reported the aorta appeared aneurysmatic in the thoracic tract, especially in the root and ascending aorta, as its axial diameter was about 6×7 cm at CT, suggesting an aortic insufficiency.

Moreover, all the aortic extension was characterized by an intimal flap indicating a type A dissection (Stanford classification), the most severe kind of dissection.

Therefore, the type A dissection concerned the ascending aorta, the aortic arch, and the descending aorta until the bifurcation in the common iliac arteries with the exclusion of the mesenteric artery and celiac trunk. Once the presence and the extension of dissection have been recognized, the radiologist must identify the double lumen representing the true and false lumens in CT scans. It must be considered that the true lumen, which was smaller because of the compression by the higher pressure of the false lumen, gave rise to the origin of the celiac trunk, superior mesenteric artery, and right renal artery. On the other hand, the false lumen, which was larger, presented delated enhancement, surrounded the outer curve of the arch, and gave rise to the left renal artery. Moreover, the false lumen had wedges around the true lumen (beak sign) and had a circular configuration because of the persistent systolic pressure, surrounding the true lumen. The radiologist excluded the presence of a thrombus in the first CTA performed. After

a few days, a new CT scan was requested confirming a critical clinical condition of mal-perfusion syndrome, a severe complication of aortic dissection (Crawford et al., 2016).

Therefore, the CT revealed a diffuse hypodensity of ischemic significance of the renal parenchyma bilaterally, with a few thin sectors remaining opaque; the lower mesenteric artery was not visible. Hepatomegaly with diffuse hepatic hypodensity was present: the over-hepatic veins were narrow but opacified and the portal vein was thin. The spleen and pancreatic tail were similarly hypodense, and there was a lot of fluid in the belly.

Unfortunately, the patient's systemic condition fell rapidly, and the patient died a few days later.

Since effective TOF repair allows many patients to live to adulthood, the population of patients with repaired TOF is rapidly growing. Nevertheless, because of the possibility of late complications, such as aortic root dilatation, these patients will require lifelong monitoring (Seki et al., 2016; Egbe et al., 2019). While aortic root dilatation is a well-known issue following TOF surgery, dissection is uncommon and maybe sometimes underreported; in fact, late aortic dissection is a rare but serious complication associated with cardiac surgery that could lead to mal-perfusion and death. The reported case suggests the importance of meticulously monitoring the patient's condition after corrective surgery for TOF.

Conclusion

In the Fallot tetralogy-treated population is required a lifetime follow-up after the radical correction and early indication for surgery for residual findings such as resolution of pulmonary regurgitation or dilatation of the aortic root is essential. Preventive surgery of the dilated aortic root will prevent aortic dissection which can be fatal for the patient.

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