Acute Coronary Syndrome in a 60 Year Old Patient with Uncorrected Tetralogy of Fallot

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Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect. Sixty-six percent of children with TOF survive the first year of life, while only 3% pass the age of 60 years without correction. The preferred age for surgical repair is 3–6 months. Only 11 cases of uncorrected TOF patients beyond the age of 60 have been described in the literature, the oldest being 87 years old. Even rarer are cases of acute coronary syndrome in patients who have not undergone surgical repair, since they usually do not survive to an age old enough to develop atherosclerosis with significant narrowing of coronary arteries.

In this paper we present the case of a 60 year old patient with uncorrected TOF who was hospitalized due to acute coronary syndrome. During catheterization significant stenosis in his right coronary artery (RCA) was demonstrated and a bare metal stent was inserted. In addition, an anomalous origin of the left descending artery (LDA) from the RCA was seen, an anomaly that characterizes about 10% of patients with TOF. In light of the patient's general condition, age and personal wish, no surgical repair was performed and the patient was discharged for further ambulatory follow-up.

PATIENT DESCRIPTION

A 60 year old Caucasian male who immigrated to Israel from the former Soviet Union in 1994 presented to our emergency department with unstable angina. His medical history included hyperlipidemia, active smoking of 10 pack-years, and unrepaired tetralogy of Fallot. Physical examination demonstrated clubbing of fingers and toes, slightly shifted cardiac apex, systolic thrill along the left sternal border, regular heart sounds with a fixed S2 split, and a long mid-systolic murmur was audible at all sites with irradiation to both carotids and left axilla. Electrocardiogram showed sinus rhythm 93/min, right axis deviation on limb leads, and poor R wave progression in V1-V4. All laboratory values were within normal range except for troponin T (0.18 ng/ml). Also notable was the hemoglobin level, 17.2 mg/dl, and O_2 saturation of 94% in room air. Chest X-ray demonstrated an enlarged heart with a boot-shaped silhouette (small pulmonary artery). Upon admission to the intensive coronary care unit an echocardiogram demonstrated preserved systolic function with a hypokinetic infero-basal segment, one large ventricular septal defect (VSD) 34 mm in diameter in the sub-aortic region, and another smaller muscular VSD in the mid-part of the interventricular septum, over-riding aorta with a slight dilatation of its root, right ventricular hypertrophy with normal size and contraction, and right ventricular outlet (RVOT) obstruction with a maximum pressure gradient of 92 mmHg [Figure 1 C-F].

Two years earlier, the patient was admitted to our cardiology department due to unstable angina. Cardiac computed tomography-angiography (CTA) showed the findings described above [Figure 1A and B], normal pulmonary arteries, and mild coronary artery calcification with anomalous origin of the left descending coronary artery from the right coronary artery. In light of these findings no invasive intervention was performed and the patient was released for further ambulatory follow-up. During the present admission, coronary catheterization was performed, which demonstrated the anomalous origin of the LAD from the RCA with anterior course to the aorta with slight irregularities along its course. A 90% stenosis in the mid-RCA was treated with bare metal stent and another 90% stenosis in the proximal part of the posterior descending artery was treated with balloon dilatation. Two days later the patient was discharged.

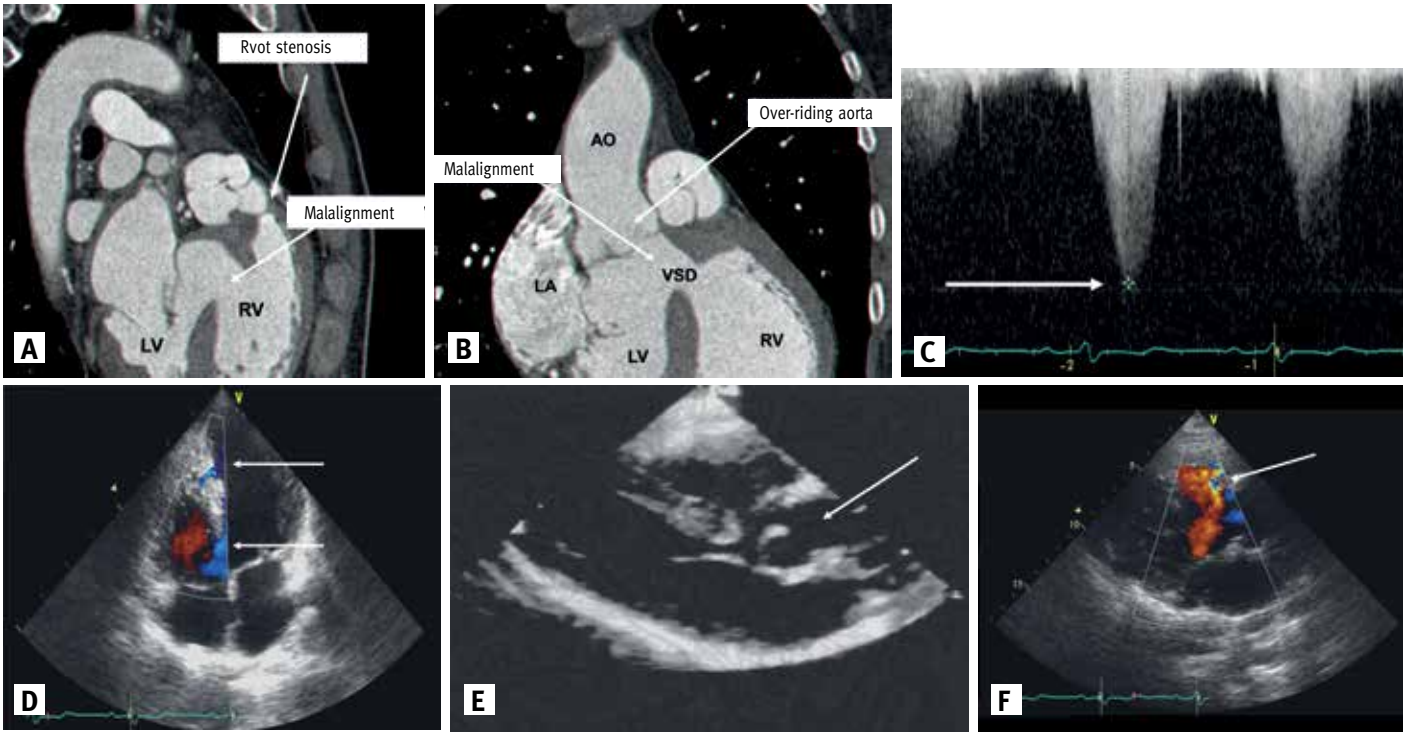
COMMENT

TOF is the most common cyanotic congenital heart defect [1], representing about 7–10% of all congenital heart defects, 3.9 per 10,000 live births, with no difference between the sexes [2]. Pathologically, TOF consists of four components:

- Ventricular septal defect, usually located in the peri-membranous part of the septum but sometimes also involving the muscular part as well. In a small number of cases there may be several VSDs.
- Over-riding aorta
- Pulmonic obstruction which can be valvular, sub-valvular, supra-valvular or mixed
- Hypertrophy of the right ventricle.

From the embryologic point of view, the cause of TOF is poor development of the spiral septum. This septum divides the truncus arteriosus during prenatal life into two vessels: the aorta and the pulmonary arteries. It then connects to the interventricular septum to complete the separation of the heart into left and right ventricles.

Figure 1. [A and B] Cardiac CT scan showing a large VSD with the over-riding aorta and RVOT stenosis, and **[C]** a very high flow velocity upon the RVOT (just over 4 m/s). **[D]** Four-chamber view showing both the septal and sub-aortic VSDs. **[E and F]** Long axis parasternal views of the over-riding aorta



If this spiral septum malaligns right to the midline, the end result will be an unusually large aorta, pulmonic stenosis and/or narrowing of the right ventricle outlet, and an imperfect connection between the two septa which leads to interventricular septal defect. Hypertrophy of the right ventricle is therefore the result of the RVOT obstruction and the left-to-right shunt caused by the VSD.

In addition to the anomalies that constitute the tetralogy itself, other abnormalities can be found in these patients, such as right aortic arch in about 25% [3] and anomalous origin of the coronary arteries in about 10% (LAD origin from the RCA is the most common anomaly in this group) [4]. Atrial septal defect (also known as the pentalogy of Fallot) is observed in about 10% of patients [3]. Repair is usually undertaken during the first year of life, best performed at age 3–6 months. Without surgical repair, these patients usually have a particularly short life expectancy: 66% survival in the first year of life; 6% will live to the age of 30, and only 3% will reach the age of 40 due to the increased risk of heart failure, endocarditis and brain abscesses [5]. The development of signs and symptoms depends on the severity of the RVOT obstruction and the size of the VSD. The first sign is usually peripheral cyanosis due to a right-to-left shunt in the heart. These children also suffer from spells of dyspnea, accompanied by very low oxygen saturation, and cyanosis that can be triggered by crying, sucking, fever or exercise. During these spells these children tend to squat, an action that raises the systemic blood pressure and thus more blood flow to the pulmonary system. As a result, they also have difficulty eating and performing routine activities and are characterized by poor growth and development. A mid-systolic murmur may be heard during physical examination, which derives from the narrowing of the RVOT but not the VSD (since the pressure gradient between the ventricles is negligible). During the cyanotic spells this murmur may disappear due to worsening in the degree of RVOT obstruction. The relative impedance between the systemic and the pulmonary circulation will ultimately dictate the clinical presentation. A large VSD with equilibration of the pressure in both ventricles and a moderate degree of RVOT obstruction that allows normal pulmonary flow will cause a variation called "acyanotic Fallot" that has no significant right-to-left shunt. Consequently, there is no systemic desaturation (normal SO_2), no pulmonary overflow and, therefore, normal pulmonary artery pressure (almost nil shunts). In the case presented here, there was marked clubbing of both hands and feet despite oxygen saturation in the normal range (acyanotic). These findings could be due to a larger right-to-left shunt during the first years of life, when the patient had normal blood pressure, causing cyanosis and clubbing. Over time, as the blood pressure increased (due to normal and pathological processes), the relative impedance between the pulmonary and systemic circulation decreased, reducing the degree of the shunt and leading to

the development of an acyanotic variant with residual clubbing.

A review of the literature disclosed 11 case reports of patients with TOF who lived beyond the age of 60 without correction, the oldest being 87 years old. Even harder to find were patients with uncorrected TOF and acute coronary syndrome since they generally do not live long enough to develop significant atherosclerosis. In our patient, the degree of RVOT obstruction prevents pulmonary overflow; therefore, pulmonary pressure is normal without pulmonary congestion, and the right ventricle contraction is still preserved so there is no systemic venous congestion. At the same time the large VSD equilibrates almost the same pressure in both ventricles with minimal bidirectional shunts and, therefore, almost nil right-to-left shunt (acyanotic). In view of the patient's condition, his age and his personal wish, no surgical intervention to correct the anomaly was performed and he was discharged for further follow-up at the adult congenital heart disease clinic.

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