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CASE REPORT

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Aberrant right coronary artery occlusion during the percutaneous pulmonary trunk stenting in a patient with tetralogy of Fallot

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Abstract Aberrant coronary arteries are frequently observed in patients presenting with Fallot's tetralogy (TOF). Before the complete surgical repair of the TOF, the percutaneously performed pulmonary trunk (PT) angioplasty is often performed in order to temporarily increase the pulmonary circulation, thus increasing the pulmonary vessel size, finally improving surgical outcome. This case reports a 12-year-old boy with a TOF insufficiently improved by surgical correction, in whom a PT angioplasty with stent implantation was complicated by an extrinsic compression of an aberrant right coronary artery (RCA) causing a myocardial ischemia with severe hypotension. The RCA, originating from the left anterior descending coronary artery, passed through the aortic root and the PT and was thus compressed by the PT-stent. Finally the RCA was successfully treated with standard coronary balloon angioplasty and stenting, improving myocardial perfusion and the hemodynamics of the patient, who finally died several days thereafter due to septic shock and massive pulmonary embolism.

Key words Aberrant coronary occlusion · Pulmonary stenting · Tetralogy of Fallot

Introduction

Tetralogy of Fallot (TOF) is one of the most observed cardiac malformations in pediatric cardiac catheterization laboratories. This cardiac malformation is also often associated with several coronary anomalies (e.g., aberrant origin), occurring in up to 7% of all TOF.[1] The most frequently observed coronary abnormality is the presence of the left anterior descending artery (LAD) or the left circumflex artery (LCX) taking off from the right coronary sinus or directly from the right coronary artery (RCA) (LAD: 37% of the cases; LCX: 26%), followed by the presence of a single coronary artery originating from the left coronary sinus (up to 15%). Other TOF-related coronary malformations are rare. Surgical correction of TOF should be attempted in all cases before adolescence, in order to avoid irreversible right heart failure, but ideally it should be performed in the first 2 years of life.

To facilitate the complete TOF surgical correction, thus improving patient outcomes, a presurgical right ventricular outflow tract (RVOT) balloon angioplasty may be attempted in order to increase the pulmonary ring and consequently the pulmonary arteries' sizes, finally correctly preparing the pulmonary circulation to the definitive surgical repair.[2,3] This percutaneous RVOT balloon angioplasty is a safe and technically relatively easy procedure; however, in this instance we report an unexpected life-threatening coronary complication that occurred during the stenting procedure of the pulmonary trunk (PT) in a patient presenting with a TOF insufficiently improved by a previously performed surgical correction.

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Case report

A 12-year-old boy from Madagascar was transferred to our tertiary center by a humanitarian organization for the cardiological treatment of a recently diagnosed TOF. Despite cyanosis being present since birth, the diagnosis of TOF was performed only when the boy was 12 years old, presenting severe dyspnea (New York Heart Association IV) with peripheral edemas.

A first attempt of surgical correction, with a pericardial patch at the ventricular septal defect (VSD) and the pulmonary supra-annular levels, was successfully performed. Further, a pulmonary valve commissurotomy with an infundibular enlargement was also performed at the same moment. Despite this successful surgical correction, 2 months later symptoms persisted, thus a new cardiological workup showed the presence of a significant post-valvular

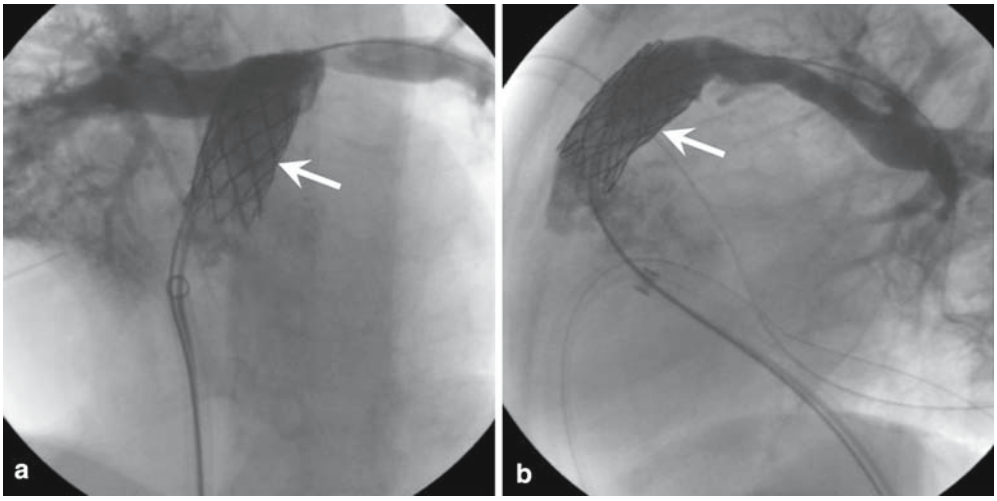

Fig. 1. **a** Frontal view of the pulmonary trunk (PT) after stenting (*arrow*) showing no significant vessel narrowing. **b** Lateral view of the PT after stenting (*arrow*) showing no significant vessel narrowing

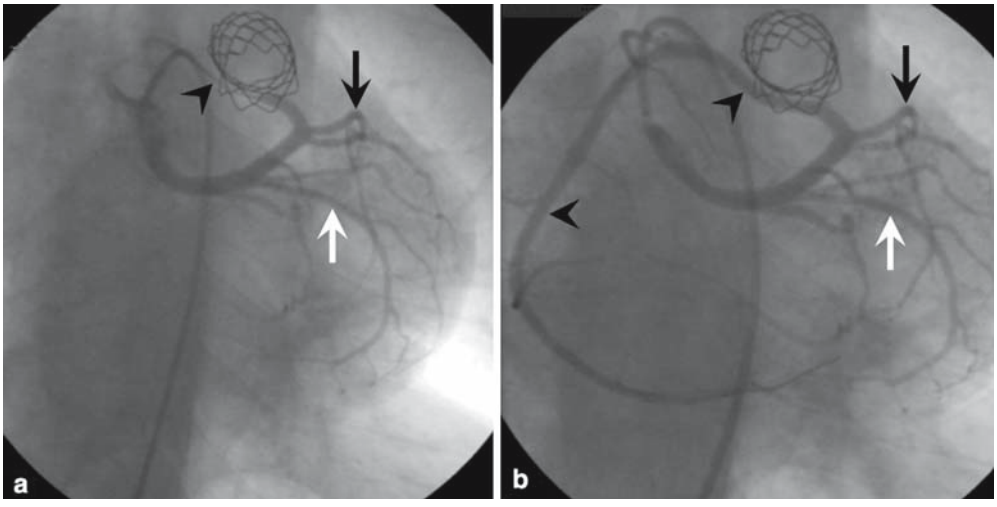

Fig. 2. **a** Selective angiography of the left main coronary showing the circumflex artery (*white arrow*), the left anterior descending (*black arrow*), and the aberrant right coronary artery (RCA) extrinsically compressed by the stent of the PT (*arrowhead*). **b** Final result after stenting of the RCA (*arrowheads*) showing no significant coronary vessel narrowing

pulmonary stenosis, which was decided to be treated by a percutaneous supravalvular stent.

Procedure

Under general anesthesia a 6-F introducer is positioned in the right common femoral vein. The main pulmonary trunk was catheterized and thanks to a 0.035-inch exchange wire, a balloon expandable NuMED CP Stent of 34-mm long (NuMED, Hopkinton, NY, USA) was advanced and finally successfully expanded by a 16-mm NuMED Balloon in Balloon (BIB: NuMED) (Fig. 1a,b). A few minutes after the stent deployment the patient became hemodynamically unstable with several episodes of bradycardia and severe hypotension. A mechanical complication (e.g., RV perforation, pulmonary artery rupture) caused by the stenting procedure was excluded by a RV ventriculography which did not show any contrast medium extravasation, and which also confirmed the good position and patency of the previously implanted stent.

Subsequently a ST elevation on the inferior leads of the surface ECG appeared, therefore we decided to perform an arterial catheterization with an ascendant aortography, in order to nonselectively check the coronary arteries status. Because the RCA was not visualized arising from the right coronary sinus, we performed a selective left coronary angiography to look for an eventual RCA aberrant origin. With a JL 3.5 6-F we confirmed the presence of a single coronary artery which normally generates the LAD and LCX, but also gives the origin of aberrant RCA coming from the LAD. The mid portion of this aberrant RCA was subocculded with a slow flow (Figs. 2a and 3a). Thanks to two orthogonal views of this aberrant RCA we were able to affirm that its trajectory was between the aortic root and the main pulmonary trunk. At this site it was clearly noted that the recently implanted pulmonary stent extrinsically compressed the aberrant RCA, mimicking an inferior ST elevation myocardial infarction (hypotension, bradycardia, ECG modifications). Due to the urgent situation we decided to attempt a coronary angioplasty of the occluded RCA. A JL 3.5 guiding catheter (Launcher Medtronic Vascular,

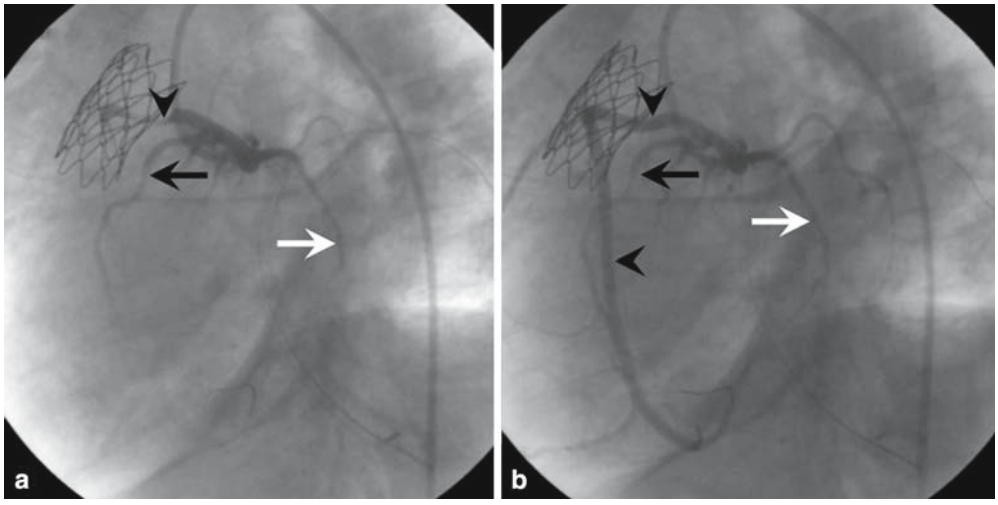

Fig. 3. **a** Left oblique view. Selective angiography of the left main coronary showing the circumflex artery (*white arrow*), the left anterior descending (*black arrow*), and the aberrant right coronary artery (RCA) extrinsically compressed by the stent of the PT (*arrowhead*). **b** Final result after stenting of the RCA (*arrowheads*) showing no significant coronary vessel narrowing

Santa Rosa, CA, USA) was selectively introduced in the ostium of the single coronary artery. A 0.014-inch BMW guidewire (Abbott Vascular, Abbott Park, IL, USA) easily passed the RCA subocclusion and a Maverick balloon 2.5/20 mm (Boston Scientific, Natick, MA, USA) at 12 atm dilated the narrowing. Due to the extrinsic compression an immediate recoil phenomenon of the lesion was observed, therefore we decided to stent the lesion with a bare metal stent offering a good radial force (Vision 3.0/20 mm at 14 atm, Abbott Vascular). Finally a good angiographic result with a normal coronary flow (Figs. 2b and 3b), and a re-establishment of the ECG modifications and of the hemodynamic condition was obtained.

Despite the successful pulmonary trunk stenting, the patient's condition in the following days further deteriorated, and 12 days thereafter he died due to neurological complications and *Pseudomonas* pneumonia complicated by a septic shock. The autopsy showed that the cause of death was a massive pulmonary embolism accompanied by a subtotal thrombotic occlusion of the previously implanted pulmonary stent.

Discussion

Fallot's tetralogy is quite often associated with coronary artery anomalies, especially aberrant origins.[1] To avoid several intraoperative surgical complications, it is important to recognize early the presence of such aberrant coronaries, in order to reduce the surgically related morbidity. Several centers regularly perform a preoperative coronary angiography to all TOF patients; however, less invasive diagnostic tools (e.g., echocardiography) are also utilized to detect such anomalies.[4]

Other noninvasive diagnostic tools (e.g., cardiac computed tomography scan and magnetic resonance imaging [MRI]) are also nowadays at the cardiologists' disposal. However, due to the rapid heart rate of these young patients a good imaging interpretation remains quite challenging.[5–7] Usually, to diminish the surgical morbidity the complete TOF surgical correction is delayed for several years, in order to previously perform a percutaneous pulmonary trunk angioplasty, aimed to let the infant grow and to allow the pulmonary arteries to sufficiently increase their caliber, thus better tolerating the final surgical correction.[2,3]

The pulmonary trunk angioplasty is relatively safe and easy to perform, and was clearly associated with a better outcome of the final complete surgical correction.[2,3] In our case an image of the coronary vessel was never obtained, because the patient was referred to our center directly from Africa in an already bad condition. Therefore, the presence of this aberrant RCA was not known by the cardiology who intended to directly stent the PT, and was finally suddenly faced, during the procedure, with this life-threatening RCA occlusion.

Due to the inherent limitation of the coronary angiography (only bidimensional evaluation of the coronary trajectory), it is often very difficult to correctly analyze the anatomy of such aberrant coronary at angiography (i.e., retroaortic trajectory, between the aortic root and the main pulmonary trunk etc.); therefore, to avoid such a complication it is recommended to perform preprocedurally a non-invasive coronary imaging able to correctly analyze in three dimensions the aberrant coronary anatomy (e.g., multislice coronary scan or MRI) even if such an image needs further improvement with the presently available technologies.[5–7]

In case of a preprocedurally detected aberrant anatomy of one of the coronary arteries, interventionists may predict the potential effect of the PT stent implantation on such an artery by placing an appropriately sized angioplasty balloon in the PT narrowing and selectively perform a coronary angiography before and while the PT balloon is inflated. In our case, the PT stent was implanted, and only after that the selective coronary angiography confirmed the aberrant RCA anatomy passing between the aortic root and the main PT. As expected, the simple balloon RCA angioplasty was insufficient to re-establish a normal coronary flow, due to the immediate recoil phenomenon caused by the pulmonary stent. Fortunately, a coronary stent with a good radial force allowed reopening of the RCA, counterbalancing this extrinsic compression.

There are very few reports in the literature on coronary artery stenting in children. However, cardiologists suggest that coronary artery lesions should be treated in the same way as in adult patients (i.e., angioplasty +/– stenting), and so far the results of small series of patients are quite encouraging.[8,9]

Our stenting strategy on this 12 year-old boy was also corroborated by the fact that the chosen stent size of 3 mm was an acceptable size for a midportion of an adult RCA, thus probably not causing any undersizing even during the future growth of the boy.

Conclusion

This case suggests that in case of TOF a preoperative (invasive or noninvasive) imaging of the coronary anatomy is mandatory, due to the increased high prevalence of aberrant coronary arteries associated with the TOF. If not performed, the interventionist, before performing the scheduled pulmonary trunk angioplasty, should inject both coronary arteries in order to detect some anomalies, thus anticipating serious complications as occurred in our case. Furthermore, in case of extrinsic coronary vessel compression caused by the pulmonary stent, standard angioplasty plus the correct choice of stenting strategy can be safely and efficaciously performed.

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