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CASE REPORT
Adult congenital heart disease

First diagnosis of severe coarctation of the aorta necessitating percutaneous intervention during pregnancy: a case report

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Background Coarctation of the aorta (CoA) is a common congenital heart defect that affects about 3–4 in every 10 000 live births. Despite clear signs on clinical examination, the diagnosis is sometimes not made until adulthood. An increasing number of patients with CoA are reaching child-bearing age. Unrepaired CoA or severe recurrent stenosis during pregnancy is a significant concern, as it poses a high risk of maternal and foetal complications and even death.

Case summary A 21-year-old woman was referred to the cardiology department at 17 weeks' gestation for management of arterial hypertension and unexplained systolic murmur. She had been diagnosed with hypertension elsewhere the year before presentation, but unfortunately, this remained unexplored. She had been started on labetalol early in the pregnancy. Clinical examination showed a loud systolic heart murmur extending from parasternal to subclavicular and scapular areas. Pulses in the lower extremities were very weak, and blood pressure was slightly elevated with a significant gradient between the upper and lower extremities. Echocardiography showed remarkable absence of pulsatile flow in the abdominal aorta and narrowing just distal to the subclavian artery with typical diastolic tail pattern on suprasternal imaging. Cardiac magnetic resonance confirmed the presence of a severe coarctation distal to the subclavian artery and presence of multiple collaterals allocating this patient in the extremely high-risk category with a risk of up to 40%–100% of maternal cardiac event during pregnancy. An extensive multidisciplinary team meeting was convened. After initial medical optimization, increased claudication and signs of placental hypoperfusion necessitated an endovascular procedure under general anaesthesia at 23 weeks' gestation. A Bentley BeGraft Plus stent (16 × 38 mm) was successfully placed. Postoperative ultrasound showed biphasic placental perfusion and normalization of blood pressure and ankle-brachial indices. At 36 weeks' gestation, the patient gave birth to a healthy child.

Discussion Coarctation of the aorta should be considered in any young patient with arterial hypertension. Altered maternal haemodynamics during pregnancy resulted in severe symptomatic CoA and reduced placental flow necessitating percutaneous intervention during pregnancy. A multidisciplinary pregnancy heart team is essential for optimal treatment management in these high-risk patients.

Keywords Coarctation of the aorta (CoA) • Pregnancy • Percutaneous treatment • Multidisciplinary pregnancy heart team • Case report

ESC curriculum 2.2 Echocardiography • 9.7 Adult congenital heart disease • 9.8 Pregnancy with cardiac symptoms or disease

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Learning points

- Always take hypertension seriously in young women.
- Coarctation of the aorta can easily be diagnosed with echocardiography focusing on subcostal and suprasternal imaging.
- Multidisciplinary approach is essential to improve prognosis in this high-risk population.

Introduction

Coarctation of the aorta (CoA) is a common congenital heart disease, accounting for approximately 5% of all congenital cardiac anomalies.[1] Up to 20% of all patients with CoA are diagnosed in adult life. In adults, CoA typically presents with hypertension, lower blood pressure (BP) and delayed pulses in the lower extremities, lower extremity fatigue, and exertional dyspnoea. A growing number of patients with CoA reach child-bearing age, which can lead to serious complications during pregnancy. Pregnancy causes significant cardiovascular changes[2] that put additional stress on the affected aorta. Untreated CoA during pregnancy can have serious consequences for both the mother and the foetus, including congestive heart failure, hypertensive crisis, aortic dissection, cerebral infarction, growth retardation, and premature birth. With a simple clinical examination and echocardiographic workup, the diagnosis of CoA can be easily made. Nevertheless, this diagnosis is often overlooked in adulthood, resulting in many often avoidable complications.

This case report presents a woman who was first diagnosed with CoA during her pregnancy. Pregnancy resulted in progressive deterioration of both the mother and the foetus, leading to the need for transcatheter treatment during her pregnancy.

Summary figure

Date	Event
2 years before presentation at our centre	Arterial hypertension during visit with general practitioner, considered as stress related, left untreated and unexplored
First trimester pregnancy	Arterial hypertension, start labetalol treatment, referral to cardiologist
17 weeks' gestation	Referral to our hospital for further exploration of unexplained systolic murmur and hypertension Diagnosis of severe CoA by clinical exam and echocardiography Multidisciplinary heart team meeting Addition of nifedipine and methyldopa to medical therapy
22 weeks' gestation	Increasing complaints of claudication, unmeasurable BP in the leg, placental hypoperfusion
23 weeks' gestation	Successful percutaneous intervention with Bentley BeGraft Plus stent (16 × 38 mm). Immediate perioperative recuperation of foetal blood supply and normalization of arm and leg BP
24 weeks' gestation	Normalization BP, lowering BP drug treatment
25–34 weeks' gestation	Stable dimensions ascending aorta, normal BPs without medical treatment
36 weeks' gestation	Caesarean section for obstetric reason. Healthy boy of 3 285 g, Apgar score of 9, 10, 10
6-month follow-up	Results complete genetic analysis: no known genetic defects
2-year follow-up	Normotensive, stable dimensions ascending aorta, well-functioning CoA stent

Case presentation

A 21-year-old patient was referred at 17 weeks' gestation to the department of congenital cardiology for further workup of persistent hypertension and an unexplained systolic murmur. Treatment with labetalol 100 mg three times a day and aspirin 80 mg once a day was initiated. Hypertensive BP had already been diagnosed before, but was left unexplored. The patient did not experience any cardiac symptoms; however, physical activity level was low. Clinical examination showed a mild combined systolic and diastolic hypertension of 145/100 mmHg in both upper limbs. The BP was significantly lower in the legs, with values of 128/78 mmHg. Cardiac auscultation revealed a loud holosystolic murmur at the level of intercostal 2, grade 3/6, extending to the subclavicular and subscapular region. Palpation of the femoral artery and posterior tibial artery revealed weak pulses.

Electrocardiogram showed a regular sinus rhythm and no signs of left ventricular hypertrophy or ischaemia. Congenital echocardiographic exam revealed significantly diminished and monophasic flow at the level of the abdominal aorta (*Figure 1A*). Suprasternal imaging confirmed the presence of CoA (*Figure 1C* and *D*). In addition, the presence of a bicuspid aortic valve and a mild dilatation of the ascending aorta of 40 mm (27 mm corrected for BSA) was demonstrated (*Figure 1E* and *F*). The bicuspid aortic valve was well functioning with no stenosis or regurgitation. Left ventricular analysis showed normal systolic and diastolic function and no signs of hypertrophy (*Figure 1B*). Cardiac magnetic resonance confirmed the presence of a severe coarctation distal from the subclavian artery, presence of multiple collaterals, and aneurysmal dilatation of the ascending aorta (*Figure 2*).

Familial history revealed aortic valve surgery at young age in the paternal grandfather. No other relatives with aortic pathology, congenital heart defect, or underlying genetic disorder were known.

Taken together, we diagnosed the presence of a severe coarctation, a bicuspid aortic valve, and a mild aneurysmal dilatation of the ascending aorta in a 21-year-old woman at 17 weeks' gestation.

Following the 2018 ESC guidelines, our patient was categorized as mWHO IV, the category of extremely high-risk pregnancy with up to 40%–100% risk of maternal cardiac event during pregnancy.[3]

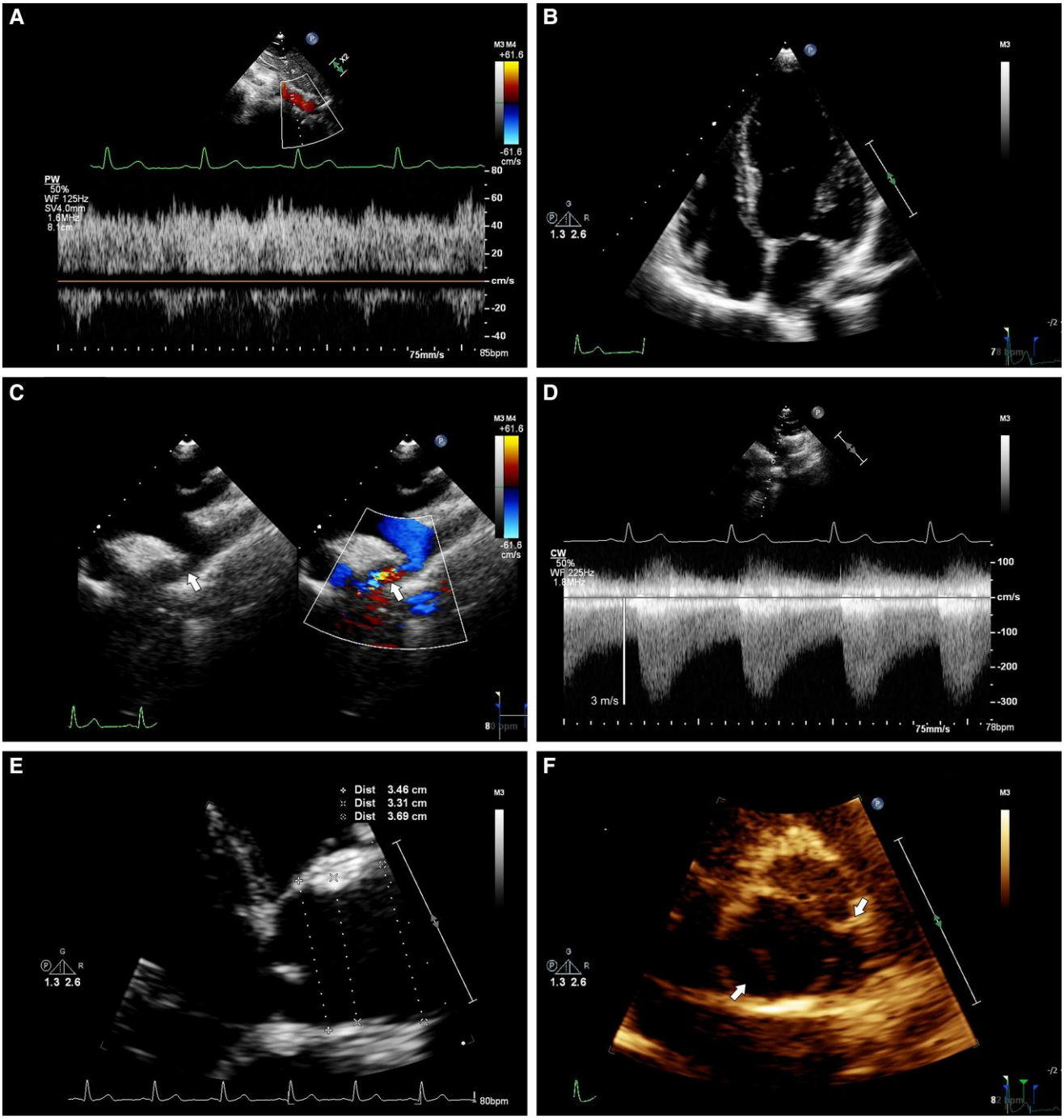

Figure 1 Baseline congenital echocardiographic exam revealed a decreased flow with monophasic signal in the abdominal aorta (*A*). Apical four-chamber view illustrated normal dimensions of the left and right heart (*B*). Suprasternal imaging showed narrowing of the descending aorta (arrow) with turbulent flow (arrow) (*C*), increased velocity up to 3 m per second (peak gradient 37 mmHg, mean gradient 15 mmHg), and a diastolic tail (*D*). Transoesophageal echocardiographic illustration of mildly dilated ascending aorta (*E*) and bicuspid aortic valve type I with the right coronary cusp (arrow below) and raphe between the left and non-coronary cusp (arrow) (*F*).

We set up an urgent multidisciplinary team meeting and organized close combined cardio-obstetric follow-up. The dedicated 20-week foetal ultrasound showed a normal morphology of the foetus, except for an abnormal flow pattern of the uterine arteries (*Figure 3A*) and a decreased growth velocity. In addition to increasing the labetalol dosage to 200 mg 3×/day, nifedipine 30 mg 1×/day and methyldopa 250 mg 1×/day were initiated as well. At 22 weeks of pregnancy, she started to complain from increased fatigue and progressive intermittent claudication in both legs. On clinical exam, BP was acceptable, but we were not able to measure the BP in the lower extremities while pulses

were extremely weak. Doppler ultrasound investigation of the lower limbs showed monophasic signals and decreased ankle-brachial indices in both legs (ABI: 0.55–0.66). Duplex ultrasound of the abdominal aorta confirmed almost no pulsatile flow while the foetal ultrasound showed increasing signs of placental hypoperfusion (*Figure 3B*).

Preliminary results of cardiogenetic screening showed a normal karyotype and no indication of (mosaic) Turner syndrome.

Regarding the increasing symptoms of bilateral intermittent leg claudication and the placental hypoperfusion, a multidisciplinary team decided to proceed with invasive treatment. At 23 weeks of pregnancy, the patient underwent an endovascular procedure under general anaesthesia.

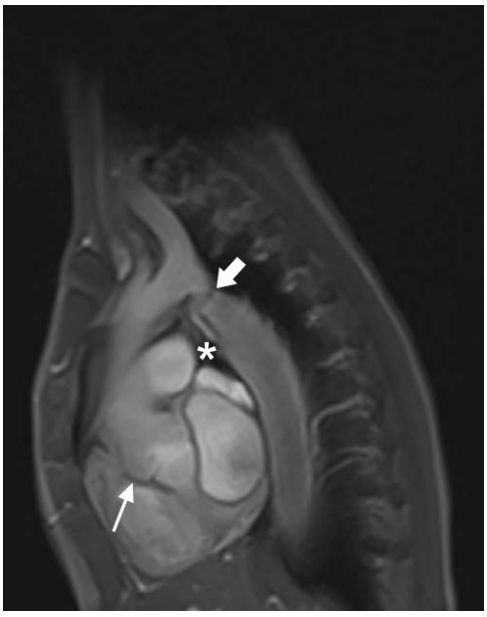

Figure 2 Cardiac magnetic resonance confirmed the presence of a severe coarctation distal from the subclavian artery (large arrow), a tortuous distal arch, and presence of multiple collaterals (*). Furthermore, the presence of an aneurysmal dilatation of the ascending aorta with diameters of 3.7 cm at the level of the aortic sinus (small arrow), 3.5 cm at the sinotubular junction, and 3.9 cm at the base of the ascending aorta was shown. Contrast was not used because of the pregnancy.

Invasive BP measurements showed a significant difference between the upper and lower extremities with almost absence of pulsatile flow in the lower extremities (*Figure 4A*). Transoesophageal echocardiography (TOE) at the level of the descending aorta revealed CoA with turbulent flow and very narrow opening (*Figure 4B*). The severe stenosis with an opening of only 3 mm was passed with a heavy-duty 0.035 mm Terumo wire. Peak-to-peak pressure gradient over the coarctation was above 30 mmHg, in line with the transthoracic echocardiographic measurements of 37 mmHg (*Figure 1D*). Next, a Bentley BeGraft Plus stent (16 × 38 mm) was successfully placed at the level of the stenosis (*Figure 4C*). Radiation exposure for this procedure with a fluoroscopic time of 2.9 min was a dose area product of 199.19 μGym^2 (19 91.1 mGycm^2) and a dose at reference point at the thoracic level of 6.9 mGy. An immediate improvement in flow pattern on TOE (*Figure 4D*), disappearance of the BP difference, and clear improvement in flow within the intrauterine arteries were all seen. The postoperative ultrasound showed normalized placental perfusion (*Figure 4E*). During cardiac follow-up, full resolution of the intermittent claudication symptoms was noted with normalization of the ankle-brachial indices on duplex ultrasound of the lower limbs. Furthermore, BP normalized and medical treatment could be stopped. During follow-up, no signs of placental hypoperfusion could be seen anymore with the foetus showing an increased growth pattern (15th centile pre-procedure vs. 70th centile at 36 weeks) (*Figure 3C*). At 36 weeks of pregnancy, the patient gave birth to her healthy son. Delivery necessitated caesarean section for obstetric reasons. Both the mother and the child are without symptoms 2 years postoperatively. Computed tomography angiography after pregnancy showed a good position of the endovascular prosthesis (*Figure 4E*).

Discussion

This case illustrates the late diagnosis of CoA in pregnancy. However, the echocardiographic diagnosis of CoA is very straightforward when focusing on the Doppler patterns in the descending and abdominal aorta. The management of CoA in pregnancy is complicated by the increased periprocedural risk to both mother and foetus. Whereas percutaneous stent implantation is minimal invasive and can result in immediate improvement of maternal cardiovascular function and placental blood flow and consequent foetal development, several risks need to be taken into account. First, aortic wall fragility due to high levels of oestrogen may lead to an increased risk of aortic dissection and rupture.[4] In addition to CoA, aortic dimensions should be closely monitored during pregnancy. Depending on the underlying condition (bicuspid aortic valve, heritable non-syndromic and syndromic disorders including Turner syndrome, Marfan syndrome, Loeys–Dietz syndrome,

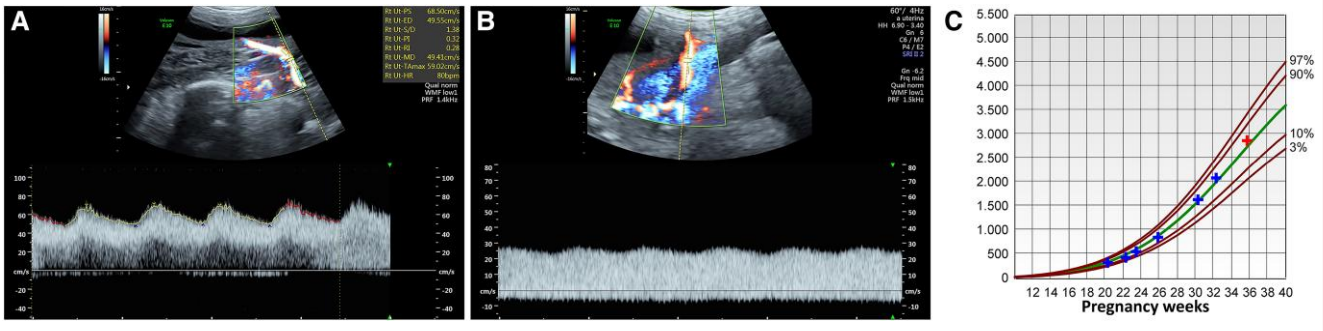

Figure 3 Pulsatility index of the uterine arteries showed placental hypoperfusion at 20weeks (PI Ut A 0.32) (*A*), with deterioration at 22 weeks to no overt biphasic flow (*B*). Foetal growth curve reveals increased foetal growth post-procedure (*C*).

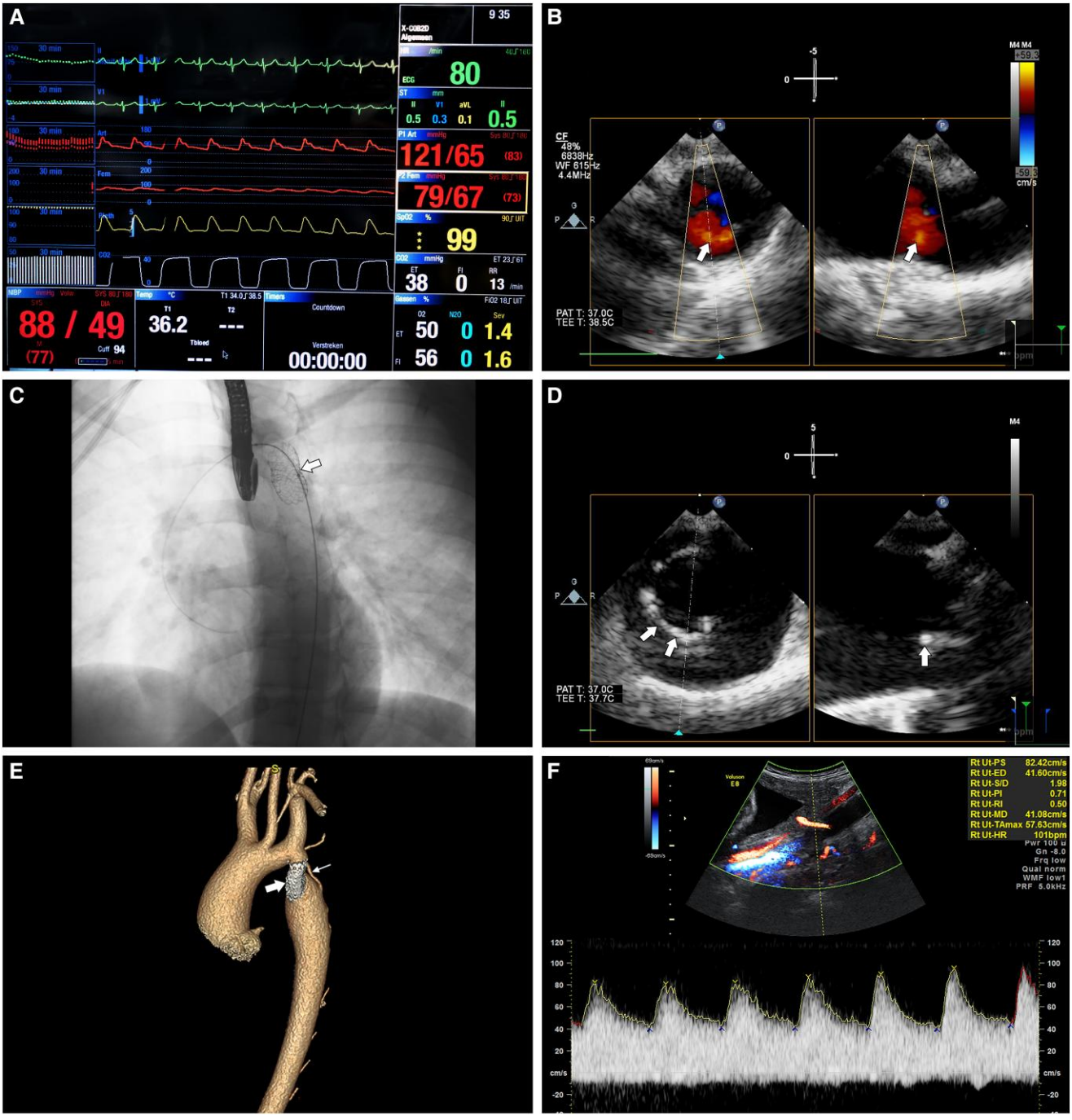

Figure 4 Imaging during and after the endovascular procedure. (*A*) Screen shot of invasive blood pressure measurements in lower and upper extremities before procedure, illustrating the significant blood pressure difference. (*B*) Transoesophageal echocardiography image of the narrow opening of the coarctation of 3 mm (arrows). (*C*) Fluoroscopic image of the stent placement (arrow). (*D*) Transoesophageal echocardiography image of the improved blood flow through the stent (arrow). (*E*) Reconstructed computed tomography angiogram of the aorta nicely shows the stent *in situ* at the site of the coarctation (large arrow) and origin of a collateral blood vessel (fine arrow). (*F*) Uterine artery flow pattern post-procedure reveals normalized placental perfusion (PI Ut A 0.71).

vascular Ehlers–Danlos syndrome, osteoaneurysm syndrome), specific aortic diameters should be considered as increased risk.[1] Second, radiation exposure radiation exposure poses a risk to the developing foetus, particularly during the first trimester when organogenesis occurs. In our case, the procedure took place in the second trimester with minimal radiation exposure focused at the thoracic level following the international recommendations.[1] Third, there is very limited data on the outcomes of percutaneous treatment of CoA during pregnancy. Finally, this procedure requires specialized material and highly skilled operators. In 2020, Ciresi *et al.*[5] presented a case of severe coarctation

in a primigravid 18-year-old woman who was successfully treated with a balloon-expanded Palmaz 3110 stent. Recently, Cherpak *et al.*[6] presented a case series of 10 patients with CoA, 4 of whom were treated percutaneously during pregnancy. However, no data on placental blood flow were presented. The pulsatility index of the uterine artery is a good measure to objectify the uteroplacental blood flow and thus the delivery of oxygen and nutrients to the foetus.[7] As presented in our case, Doppler measurements of uteroplacental flow are similar to the flow pattern in the maternal abdominal aorta. Taking into account the complexity and high-risk management of severe CoA, all patients with CoA should be followed in an expert centre with a multidisciplinary approach.

Conclusion

Coarctation of the aorta should be considered in every young patient presenting with arterial hypertension. Altered maternal haemodynamics during pregnancy resulted in severe symptomatic CoA and reduced placental flow necessitating percutaneous intervention during pregnancy. A multidisciplinary pregnancy heart team is essential for optimal treatment management in this high-risk patient.

Lead author biography

An M. Van Berendoncks is a senior staff member at the Department of Cardiology at Antwerp University Hospital with main focus on cardiac imaging, congenital heart disease, and cardio-obstetrics. She completed a fellowship in Congenital Heart Disease at the Thorax Centre, Erasmus MC, Rotterdam, under the supervision of Jolien Roos-Hesselink, and obtained the European Certificate for Congenital Echocardiography. She initiated a specialized unit for the care of ACHD patients and a pregnancy heart team at the Antwerp University Hospital, which has grown into a multidisciplinary and multicentric collaboration. She is a board member of the Belgian Working Group of Congenital Heart Disease and Belgian Working Group of Non-Invasive Cardiac Imaging.

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Consent: The authors confirm that written consent for submission and publication of this case report including the images and associated text has been obtained from the patient in line with the COPE guidelines.

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Data availability

The data underlying this article are available in the article and in its online supplementary material. If additional data are required, these data will be shared on reasonable request to the corresponding author.

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