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## **Brief Report**

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# Against all odds: successful pregnancy in an adult patient with unrepaired coarctation of the aorta

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#### Abstract

According to the modified World Health Organization classification, pregnant women with unrepaired aortic coarctation are at very high risk for both maternal and fetal complications and should, therefore, be counselled against pregnancy. The most frequent maternal complications include systemic hypertension, renal failure, preeclampsia, and aortic dissection. Herein, we describe a successful pregnancy in an adult patient with unrepaired aortic coarctation.

## **Clinical case**

According to the current guidelines,<sup>1</sup> women with unrepaired coarctation of the aorta are at increased risk for pregnancy, with various maternal complications including aortic rupture and dissection, cerebral haemorrhage due to rupture of cerebral aneurysms, congestive heart failure, hypertensive crisis, and infective endocarditis. An excess of miscarriages and hypertensive disorders has been reported as well.<sup>2</sup> Therefore, taking into account the extremely high risk of maternal mortality and severe morbidity (Modified World Health Organization Class IV), termination of pregnancy should be discussed and offered to these patients.<sup>3</sup> Herein, we describe a 24-year-old woman who presented to our Adult Congenital Heart Disease clinic at 14 weeks gestation, suffering from systemic hypertension. Physical examination revealed high blood pressure values (180/110 mmHg at the right upper arm) with a gradient higher than 20 mmHg between upper and lower extremities, as well as poor femoral pulses. The echocardiographic examination showed severe aortic coarctation with a peak gradient of about 80 mmHg at the level of the aortic isthmus, a diastolic run-off, a fixed Doppler profile of the abdominal aorta, and a mildly dilated ascending aorta (Figs. 1 and 2).

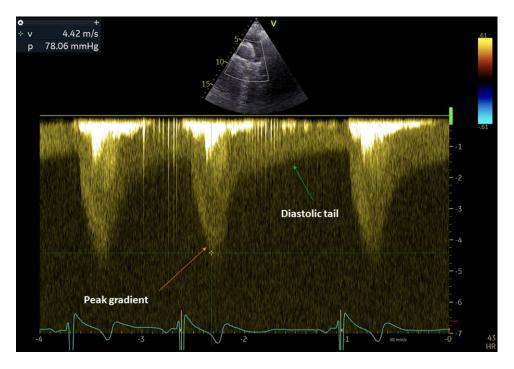
According to the current guidelines, the patient was considered to be at extremely high risk for both maternal and fetal mortality and morbidity, and, therefore, she was counselled for termination of pregnancy. Nonetheless, the patient chose not to terminate the pregnancy. We decided then to start methyldopa in order to lower the blood pressure and to perform a close follow-up to check her blood pressure and her left ventricular systolic function. The patient attended our outpatient clinic at least once a month, and screening by fetal echocardiography and a careful obstetric evaluation were performed as well. During follow-up, the size of the aorta remained stable and the left ventricular systolic function did not deteriorate; however, the blood pressure values became progressively elevated despite the increased dosage of methyldopa. Since the fetal growth was not affected and the patient remained stable, we decided not to perform any percutaneous intervention. In the end, pregnancy was successfully carried to term and a healthy newborn was delivered via cesarian section, in the absence of both maternal and fetal complications. Both labour and delivery were performed in a dedicated centre with high expertise in pregnancy and cardiac disease. The newborn was 38 weeks gestational age, the birth weight was 2.8 kilograms, and no major neonatal complications occurred.

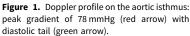
## Discussion

Unrepaired coarctation of the aorta confers a very high risk for pregnancy. Indeed, cardiovascular physiology undergoes profound changes during pregnancy: heart rate and stroke volume increase as well as cardiac output rises by 50%. Hormonal factors weaken the arterial wall and blood vessel remodelling occurs. Pregnancy-related cardiovascular changes place additional stress on the physiology of aortic coarctation. The blood pressure balance suddenly changes in the 7th month of pregnancy and, during labour, blood pressure and cardiac work rise by about 20% at the peak of each uterine contraction. In presence of systemic hypertension, a close cardiologic monitoring during gestation is therefore mandatory.<sup>1,3</sup> However, systemic hypertension must be treated not too aggressively, to avoid

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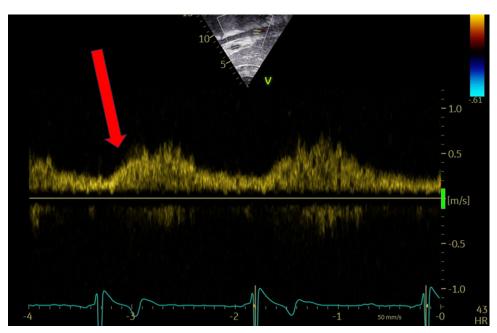


Figure 2. Doppler profile of the abdominal aorta: continuous abdominal aortic flow (red arrow).

the risk of placental hypoperfusion, and the most common drugs such as angiotensin-converting-enzyme inhibitors, angiotensin-receptor blockers, amiodarone, and spironolactone are prohibited.<sup>4–6</sup> Percutaneous treatment of aortic coarctation is feasible during pregnancy; however, since the risk of dissection is increased, it should be considered only in case of resistance to therapy as well as in case of maternal-fetal compromise.<sup>7,8</sup> Although guidelines do not exclude vaginal delivery, it is common in these patients to perform caesarean section.<sup>6</sup> Last but not least, recent studies suggest that, in women with unrepaired coarctation of the aorta, mainly due to new therapeutic advances, pregnancy may be safer than previously considered.<sup>8,9</sup>

#### Conclusion

Despite the increased risk for both maternal and fetal complications related to unrepaired coarctation of the aorta, a successful pregnancy may be possible even in women with this challenging condition. A close monitoring as well as an adequate and adjusted therapy for systemic hypertension are both paramount to carry the pregnancy to term and avoid complications.

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