A case of aortic dissection in a pregnant patient with Marfan syndrome

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Objective
To demonstrate a rare case of aortic dissection in a pregnant patient with Marfan syndrome (MFS). MFS is an autosomal dominant disease with incidence of 1:3000 – 5000 and ocular, cardiovascular and musculoskeletal complications. Pregnancy and postpartum period poses risk of aortic dissection or aortic rupture in women with MFS, due to increased arterial wall stress, caused by hypervolemic and hyperdynamic state and hormonal changes. Increased aortic root dilatation (>40mm), assessed by transthoracic ultrasound, is a predictor of aortic dissection and rupture in women with MFS (10% risk; compared to <1% risk with aortic root <40mm). Clinical picture presents with tearing and ripping sternal pain, radiating to the back. Complications of dissection, as syncope, cardiac or cerebrovascular infarction, cardiac failure, paraplegia can occur. Given the lethal nature of aortic dissection in the first 48 hours, prompt diagnosis and intervention are crucial. Due to its reliability and availability, CT is the investigation of choice, before MR and transoesophageal echocardiogram. Depending on site of dissection, aortic repair in Type A dissection (involving ascending aorta) or medical therapy (strict blood pressure control and reduction of shear stress) or percutaneous intervention in Type B (isolated descending aorta - 20%) is suggested.

Methods
This is a case report.

Results
A 30-year-old primipara with known MFS was admitted to the high risk pregnancy unit for surveillance because of dilated bulbus aortae >43mm and aortic regurgitation (WHO class III) to ensure compliance to antihypertensive therapy, avoidance of physical activity and blood pressure control. Her average blood pressure was 130/75. The patient did not comply to bed rest. On the 3rd day, after making a sudden move, she complained of a dull subscapular pain, that got worse on palpation, improved after intramuscular analgesic and was therefore evaluated as osteomuscular pain. Two days later the pain worsened and was accompanied by elevated CRP, tachypnoa, drowsiness and altered mental state. CT angiography revealed Type B aortic dissection with both lumens patent, with only right renal artery exiting from the false lumen. We administered lung maturation drugs, performed an emergency cesarean section and delivered a 1840g newborn with Apgar score 8/8. The patient was admitted to the intensive care unit with strict bed rest and blood pressure control. Subsequent CTA and MRI showed no progression of dissection. The cardiovascular surgeons agreed on conservative treatment due to limited data of successful percutaneous stent insertion and high risk of paraplegia in patients with MFS. Patient was discharged hemodynamically stable after 3 weeks, with strict physical activity restrictions and follow up in 3 months. Unfortunately, MFS was diagnosed in newborn as well.

Conclusion
Aortic dissection is a lethal condition, that necessitates prompt diagnosis and treatment. In pregnancies with MFS, aortic dissection needs to be excluded first, despite of occasionally misleading clinical presentation.

CT angiography. The arrows show sites of aortic dissection.