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ESC European Society of Cardiology

European Heart Journal - Case Reports (2021) **5**(8), 1–6
doi:10.1093/ehjcr/ytab288

CASE REPORT
Other

4D flow MRI of type B dissection with later retrograde progression to type A dissection in Marfan: a case report

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Received 28 March 2021; first decision 22 May 2021; accepted 30 June 2021

Background Due to the malfunction of connective tissue, Marfan patients are at increased risk of aortic dissection. Uncomplicated acute type B dissection is usually managed with medical therapy. Retrograde progression or new type A dissection is a relatively rare but often fatal complication that occur most frequently in the first 6 months after acute type B dissection.

Case summary We present a 31-year-old male with Marfan syndrome and a recent uncomplicated type B dissection from the left subclavian to the right common iliac artery who underwent 4D flow magnetic resonance imaging (MRI). The dissection had a large proximal intimal tear just distal to the left subclavian artery (15 mm) and large false lumen (35 mm). Aortic blood flow just distal to the left subclavian artery (3.6 L/min) was split disproportionately into the true (0.8 L/min, 22%) and false lumen (2.8 L/min, 78%). 4D flow streamlines revealed vortical flow in the proximal false lumen. Increased wall shear stress was observed at the sinotubular junction (STJ), inner wall of the ascending aorta and around the subclavian artery. Two weeks after MRI, the patient presented with jaw pain. Computed tomography showed a type A dissection with an entry tear at the STJ for which an acute valve-sparing root, ascending and arch replacement was performed.

Discussion Better risk assessment of life-threatening complications in uncomplicated type B dissections could improve treatment strategies in these patients. Our case demonstrates that besides clinical and morphological parameters, flow derived parameters could aid in improved risk assessment for retrograde progression from uncomplicated type B dissection to acute type A dissection.

Keywords Marfan • Aortic dissection • Magnetic resonance imaging • 4D flow • Case report

Learning points

- Retrograde progression or a new type A dissection is a relatively rare, but often lethal complication that occurs most frequently in the first 6 months after acute type B dissection.
- Magnetic resonance imaging 4D flow derived parameters such as elevated wall shear stress, false lumen flow volume, and vortical flow could aid in improved risk assessment for retrograde progression from uncomplicated type B dissection to acute type A dissection.

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Handling Editor: Harry Klimis
Peer-reviewers: Annagrazia Cecere and Luca Arcari
Compliance Editor: Brett Sydney Bernstein
Supplementary Material Editor: Ayse Dhahit

Introduction

Marfan syndrome is a disorder of the connective tissue caused by a mutation in the FBN1 gene.[1] Aortic dissection or rupture is the most lethal complication observed in Marfan patients, however, early recognition and (pre-emptive) treatment have significantly improved life expectancy.[2] Uncomplicated acute type B aortic dissection (without malperfusion, rapid expansion, rupture, persistent pain, or uncontrolled hypertension) is usually managed with medical therapy aimed at controlling blood pressure, with 1-year survival of ∼80%.[3,4] Retrograde progression or a new type A dissection is a relatively rare but often fatal complication reported in 4–8% of acute type B dissections, with mortality rates of up to 80%.[3,5] A new type A dissection after acute type B dissection occurs most frequent in the first 6 months.[5] Few studies have investigated risk factors for a new type A dissection after acute type B dissection, with only one study reporting flow parameters.[3,5,6] They found that a diameter of >40 mm, bicuspid valve, young age, and elevated wall shear stress (WSS) distal to the subclavian artery are predictive of a type A dissection. Other studies have shown that flow parameters such as false lumen flow can predict false lumen dilatation, illustrating the added value of these parameters beyond aortic dimensions in the risk assessment of type B dissections. Although currently follow-up imaging of aortic dissection is frequently performed using computed tomography (CT), this could change in the future due to the additional information that magnetic resonance imaging (MRI) 4D flow can provide using 3D velocity-encoding to visualize not only blood flow patterns and volumes but also WSS.

We present a case of a 31-year-old male with Marfan syndrome and a recent acute type B dissection who underwent a 4D flow MRI 2 weeks before developing a type A dissection.

Timeline

2007	18 years old	Spontaneous pneumothorax treated by drainage via a chest tube, with a recurrence later that year for which video-assisted thoracoscopic surgery is performed
2008	19 years old	Patient is diagnosed with Marfan. On magnetic resonance imaging (MRI) a maximal aortic diameter of 35 mm at the level of the sinus
2020	31 years old	Progression of aortic dimensions, on echocardiography an aortic sinus of 41 mm
3 months prior to admission		
Admission date		Acute type B dissection, from left subclavian artery to the right common iliac artery
5 weeks post-admission date		MRI with 4D flow of the aorta
7 weeks post-admission date		Type A dissection with entry tear at the level of the sinotubular junction, involvement of the innominate artery, and the left common carotid artery. Acute valve-sparing root, ascending and arch replacement (David procedure) with elephant trunk was performed

Case presentation

A 31-year-old male with Marfan syndrome and a history of recurrent pneumothorax presented with back pain, nausea, and vomiting during exercise and was diagnosed on CT with an uncomplicated type B aortic dissection from the left subclavian artery to the right common iliac artery. The proximal intimal tear was located just distal to the left subclavian artery with a maximum diameter of 15 mm, and a large false lumen (max diameter 35 mm) (*Figure 1A and B* and *Video 1*). Physical examination revealed no abnormalities and electrocardiogram (ECG) was normal. The patient was treated medically with a betablocker and calcium antagonist and follow-up imaging were planned. An MRI was performed 5.5 weeks after the type B dissection, which was already scheduled due to an increase in aortic diameter at the outpatient clinic earlier that year. In our tertiary centre aortic aneurysm in young patients are routinely monitored using MRI, including aortic 4D flow analysis. The MRI showed a dilated root of 40 mm and known type B dissection. Aortic lumen was split just distal to the left subclavian artery into true and false lumen, which directed blood volume (3.6 L/min) mainly towards the false lumen (true lumen 0.8 L/min, 22% and false lumen 2.8 L/min, 78%). On 4D flow streamlines did not indicate helicity or vorticity in the ascending aorta or aortic arch, however, vorticity was observed in the proximal false lumen (*Figure 1C and D* and *Video 2*). Increased WSS was observed at the sinotubular junction (STJ), inner wall of the ascending aorta, and just proximal and distal to the subclavian artery at the location of the dissection (*Figure 2*).

Two weeks after MRI, the patient presented to the emergency department with jaw pain, visual complaints, and decreased sensibility of the right arm. On physical examination, the patient was briefly hypotensive [86/56 mmHg, after fluid challenge 117/70 mmHg (left equals right)], with good peripheral pulses and no other abnormalities. ECG was normal. CT showed a type A dissection with an entry tear at the STJ and involvement of the innominate and left common carotid artery for which an acute valve-sparing root, ascending and arch replacement (David procedure) was performed (*Figure 1E and F* and *Video 3*). During surgery the entry tear at the STJ was also observed. *Figure 3* shows the location of elevated WSS at the STJ and ascending aorta at the time of type B dissection, which seems to correspond to the future entry tear location and true/false lumen border of the type A dissection, respectively. Postoperatively, the patient suffered from transient ischaemic attacks with amaurosis fugax and transient sensibility disorders of the right arm. He was discharged after 10 days of hospitalization. At the outpatient clinic the patient showed stable

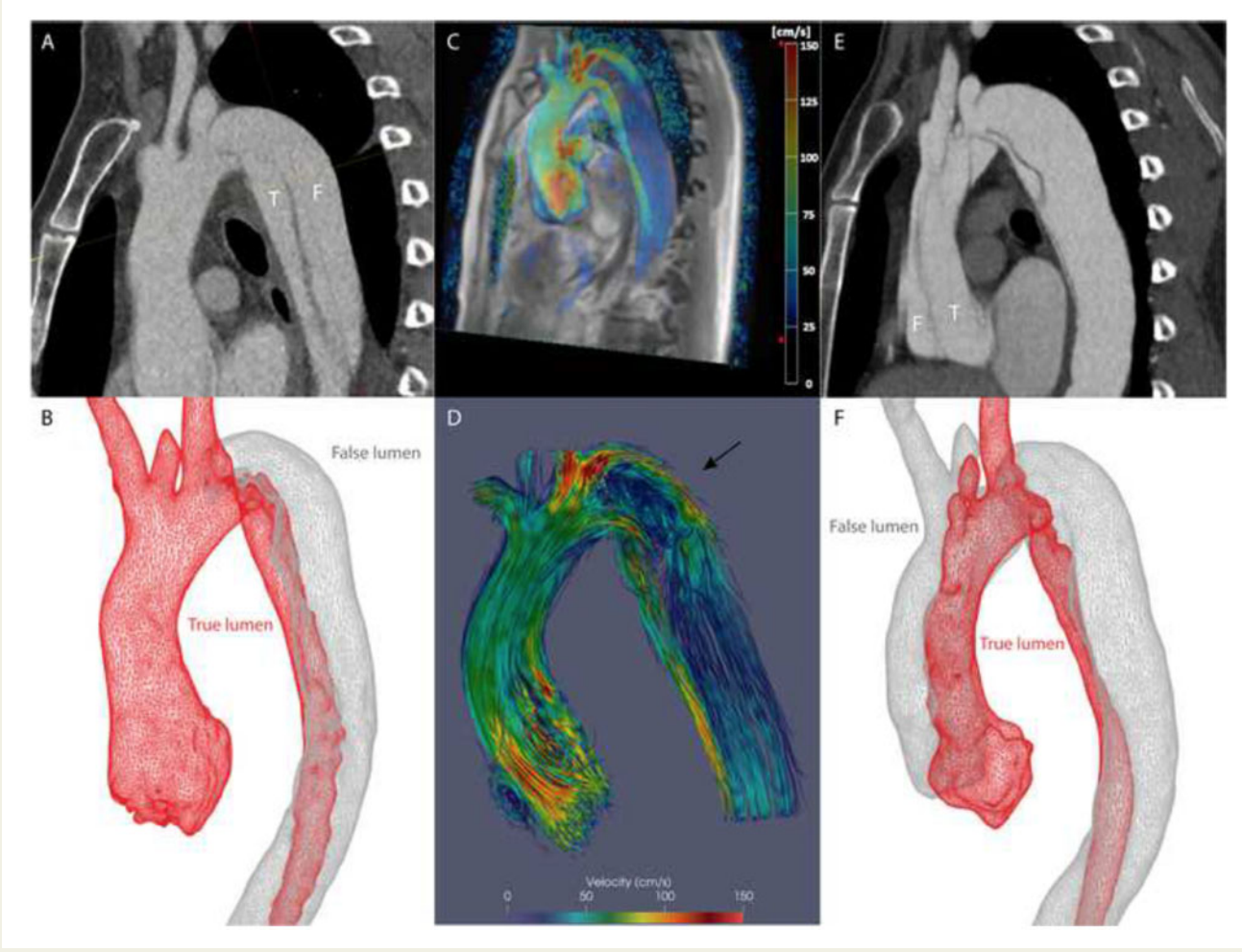


Figure 1 (*A*) T = 0: computed tomography image of type B dissection (T indicates true lumen, F indicates the false lumen) and (*B*) 3D segmentation. (*C*) $T_0 + 5.5$ weeks: 4D flow magnetic resonance imaging of type B dissection and (*D*) 3D peak-systole streamline visualization with vortical flow in the proximal false lumen (designated by the arrow). (*E*) $T_0 + 7.5$ weeks: computed tomography images of the type A dissection (T indicates true lumen, F indicates the false lumen) and (*F*) 3D segmentation. (*C* and *D*) Colour scale from blue (representing low velocity) to red (representing high velocity). Colour scale is provided next to the image.

cardiovascular parameters with no cardiac complaints. Follow-up CT imaging at 7 months showed some progression of dilatation in the descending aorta and a dissection in the left subclavian artery for which frequent imaging will be continued.

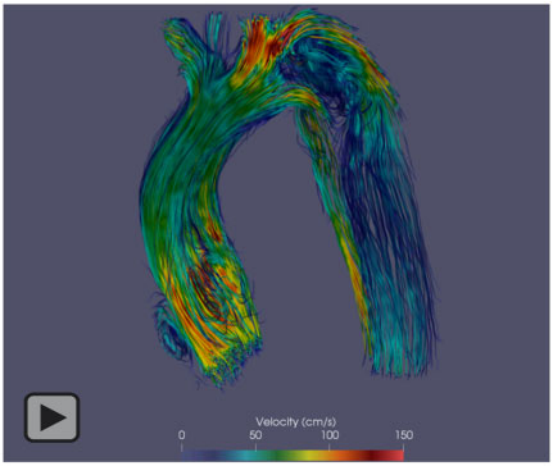
Discussion

This case presents MRI 4D flow analyses just 2 weeks prior to a relatively rare new retrograde type A aortic dissection after a recent acute type B aortic dissection.

Standard management of uncomplicated type B dissections is optimal medical treatment, however, with the evolvement of less invasive thoracic endovascular aortic repair (TEVAR), surgical interventions have become more frequent and could improve outcome in selected cases.[7] However, for Marfan patients open surgery may still be preferable as this has shown favourable outcome compared to TEVAR.[8]

To stratify who may benefit from surgical intervention, several studies have focused on finding imaging risk factors that precede adverse aortic events. For uncomplicated type B dissections a proximal entry tear of >10 mm, aortic diameter >44 mm, patent false lumen and false lumen diameter of >22 mm are predictors of adverse events, of which all except for the diameter of >44 mm were present in our case.[9–11] Furthermore, an MRI 4D flow study has demonstrated that high blood flow volume over the false lumen, in this patient 78% of total flow, is a prognostic factor for false lumen dilatation.[12] Also, vortical flow was observed in the proximal false lumen (*Figure 1D*), which has been linked to aortic dilatation in Marfan patients.[13] Flow parameters have also been assessed based on CT images combined with computational fluid dynamics, illustrating the need for more information beyond diameters.[6] They found elevated WSS just distal to the subclavian artery origin in patients with a type B dissection was associated with retrograde type A dissection, which was also observed in our case. In addition, the previous study also found that elevated

Video 1 3D segmentation of type B dissection, with true lumen in red and the false lumen in grey colour coding.



Video 2 4D flow magnetic resonance imaging peak-systole streamline visualization of type B dissection.

WSS corresponded to the site of the future dissection entry tear of a retrograde type A dissection.[6] Interestingly, in our case elevated WSS was observed at the location of the STJ which seems to correspond to the future entry tear location (*Figure 3*).

Due to the malfunction of the connective tissue, Marfan patients are at increased risk for aortic dissection as well as subsequent surgical intervention after type B dissection.[8] We hypothesize that in the type B dissection, elevated WSS at the subclavian artery and STJ, in combination with the large proximal entry tear and corresponding high volume vortical blood flow over the false lumen has led to the new type A dissection in the structurally abnormal aorta of this Marfan patient.

While no conclusion on the effect of individual parameters leading to type A dissection can be drawn from a case report, we would like to

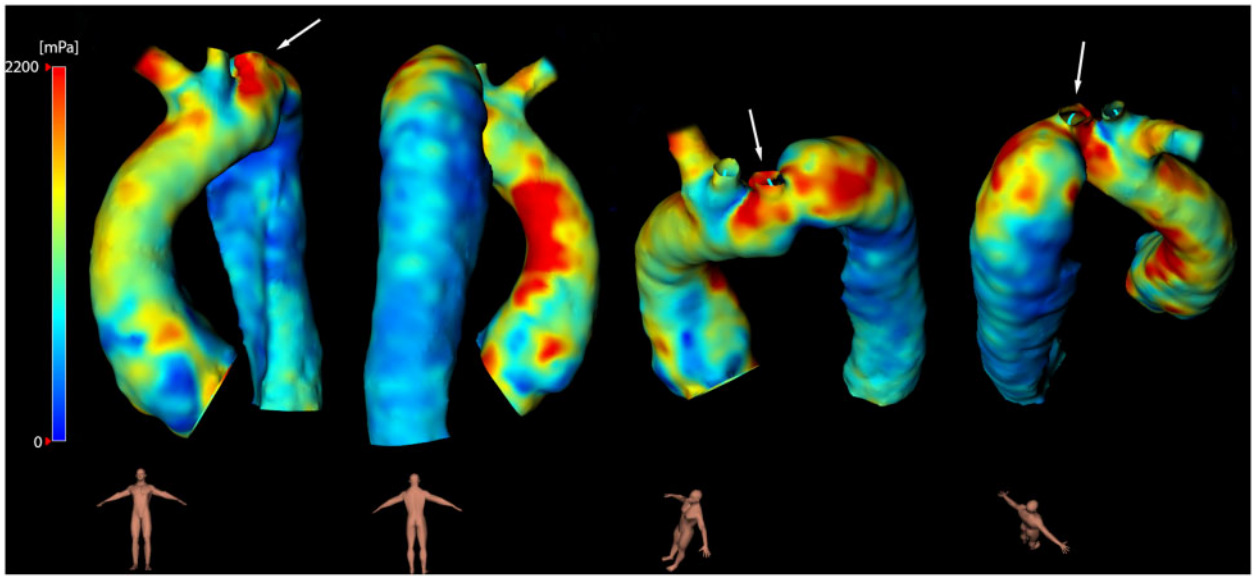


Figure 2 Type B dissection ascending, arch and false lumen (true lumen not included) wall shear stress magnitude maps derived from 4D flow magnetic resonance imaging. The arrow indicates the location of high wall shear stress surrounding the left subclavian artery. 4D flow parameters were analysed using CAAS MR Solutions 5.2; Pie Medical Imaging, Maastricht, The Netherlands. Colour scale from blue (representing low wall shear stress) to red (representing higher ≥2200 mPa wall shear stress). Colour scale is provided on the left side of the image.

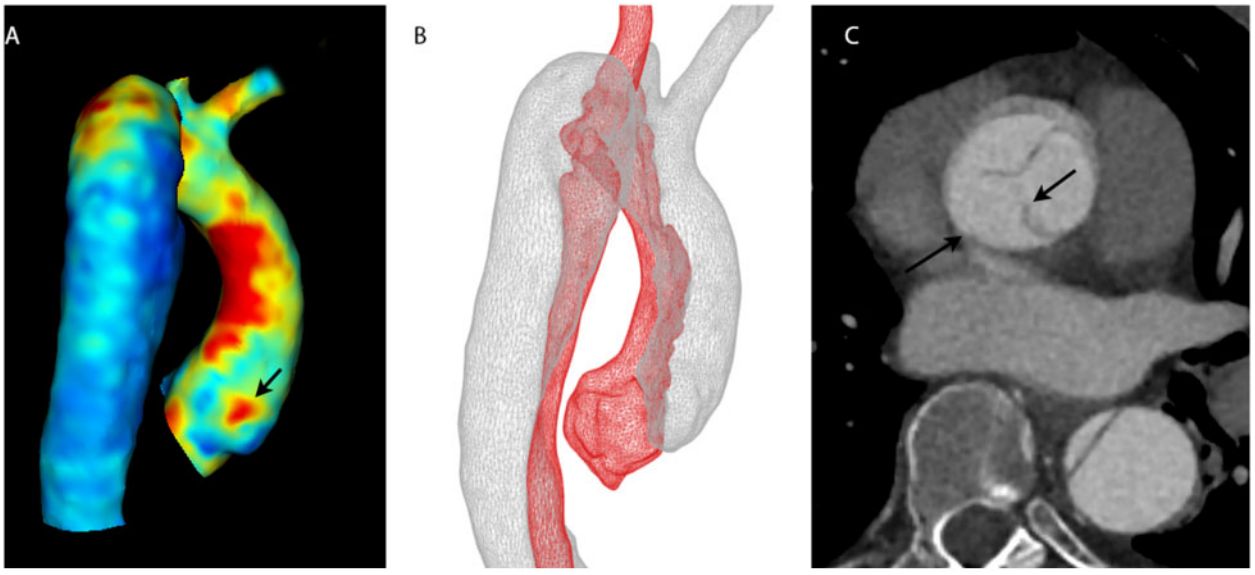


Figure 3 (*A*) Posterior view of type B dissection ascending, arch and false lumen wall shear stress. Elevated wall shear stress was observed at the sinotubular junction (indicated by the arrow) and along the inner curvature of the ascending aorta. (*B*) Posterior view of 3D segmentation of type A dissection. Please note elevated wall shear stress at the inner curvature of the ascending aorta (*A*) and the corresponding true/false lumen border of the type A dissection (*B*). (*C*) Axial computed tomography images of the type A dissection at the level of the sinotubular junction. Please note elevated wall shear stress at the sinotubular junction on the right side of the inner curvature of the type B dissection [designated with an arrow in (*A*)] and the corresponding suspected entry tear location (*C*). (*A*) Identical colour coding was used as in *Figure* 2; blue represents low wall shear stress and red represents higher wall shear stress (>2200 mPa). (*B*) Red colour = true lumen, grey colour = false lumen.

Video 3 3D segmentation of type A dissection, with true lumen in red and false lumen in grey colour coding.

encourage future research to investigate the predictive value of various morphological and flow derived parameters. Possibly subcategorization of uncomplicated type B dissections into high and low risk based on clinical, anatomical, as well as flow parameters could improve risk stratification and optimize treatment strategies in these patients.

Conclusion

Type A dissection is a rare but often lethal complication that occurs most frequently in the first 6 months after acute type B dissection. Besides clinical and morphological parameters, flow derived parameters such as vortical flow and elevated WSS could aid in improved risk assessment for retrograde progression from uncomplicated type B dissection to acute type A dissection.

Lead author biography

Max J.P. van Hout, MD, has almost 2 years clinical cardiology experience and is currently working on finishing his PhD focused on Magnetic Resonance Imaging of the aorta at the Leiden University Medical Center in The Netherlands. He plans on pursuing a career in clinical cardiology.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

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

Conflict of interest: None declared.

Funding: None declared.

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