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Brief Report

Fatal dissection of the descending aorta after implantation of a stent in a 19-year-old female with Turner's syndrome

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Abstract We report a fatal dissection of the descending aorta as a complication after a two-staged implantation of a stent to relieve aortic coarctation in a young female with Turner's syndrome. Implantation of the stent, with dilation up to 70 percent, and half a year later re-dilation to 100 percent, was without any complication. A week after the re-dilation, however, the patient suffered acute dissection of the descending aorta. We initially stabilized the situation, but some days later her condition deteriorated, and she died. We discuss the various options for treating coarctation and re-coarctation. As far as we know, this is the first description of implantation of a stent in the setting of coarctation in Turner's syndrome, albeit with an unfortunate conclusion.

Keywords: Interventional procedure; balloon angioplasty; re-dilation

Balloon angioplasty, with implantation of stents, is a well-established treatment for native coarctation in older children, adolescents, and adults, and for re-coarctation of the aorta after surgical treatment. Two-staged implantation of stents, and the use of a covered stent, have recently been advocated as therapeutic options. In this report, we describe fatal aortic dissection after a two-staged implantation of a stent in a patient with Turner's syndrome.

Case report

A 19-year-old female with Turner's syndrome was found to have an asymptomatic native aortic coarctation in the setting of a trifoliate aortic valve. Systolic blood pressure in both upper limbs was 135 millimetres of mercury, and in the lower limbs 95 millimetres of mercury (Fig. 1a). A two-staged balloon angioplasty was planned, to include implantation of a stent. So, in August 2003, we implanted a Palmaz P188 stent, dilating it to 14 millimetres, twice the diameter of the stenosis, and leaving a residual gradient of 15 millimetres of mercury (Fig. 1b). In March, 2004, she then underwent re-dilation of the stent with a balloon of 18 millimetres diameter, having easily traversed the stent (Fig. 1c). She was discharged without any complaint. One week later, she presented in the emergency room with severe chest pain of sudden onset. Transoesophageal echocardiography showed dissection of the descending aorta just below the stent. A computed tomographic scan confirmed this diagnosis, revealing true and false lumens of the descending aorta, the latter extending into the renal arteries (Fig. 1d). Therapy with antihypertensive drugs, beta-blockers and pain medication was initiated, and her condition stabilised. Two days later, however, her condition deteriorated. An additional computed tomographic scan showed further widening of the false lumen, and very poor perfusion of the kidneys was detected (Fig. 1e). The poor prognosis of this case was discussed with several colleagues from inside and outside our institution. It was judged that both surgery and endovascular repair were not options for treatment, and unfortunately she died one day later. Permission for autopsy was not granted.

Review of the family history revealed that her mother had died in November, 2001, also due to dissection of the ascending aorta. Histological investigation in the case of the mother showed fragmentation

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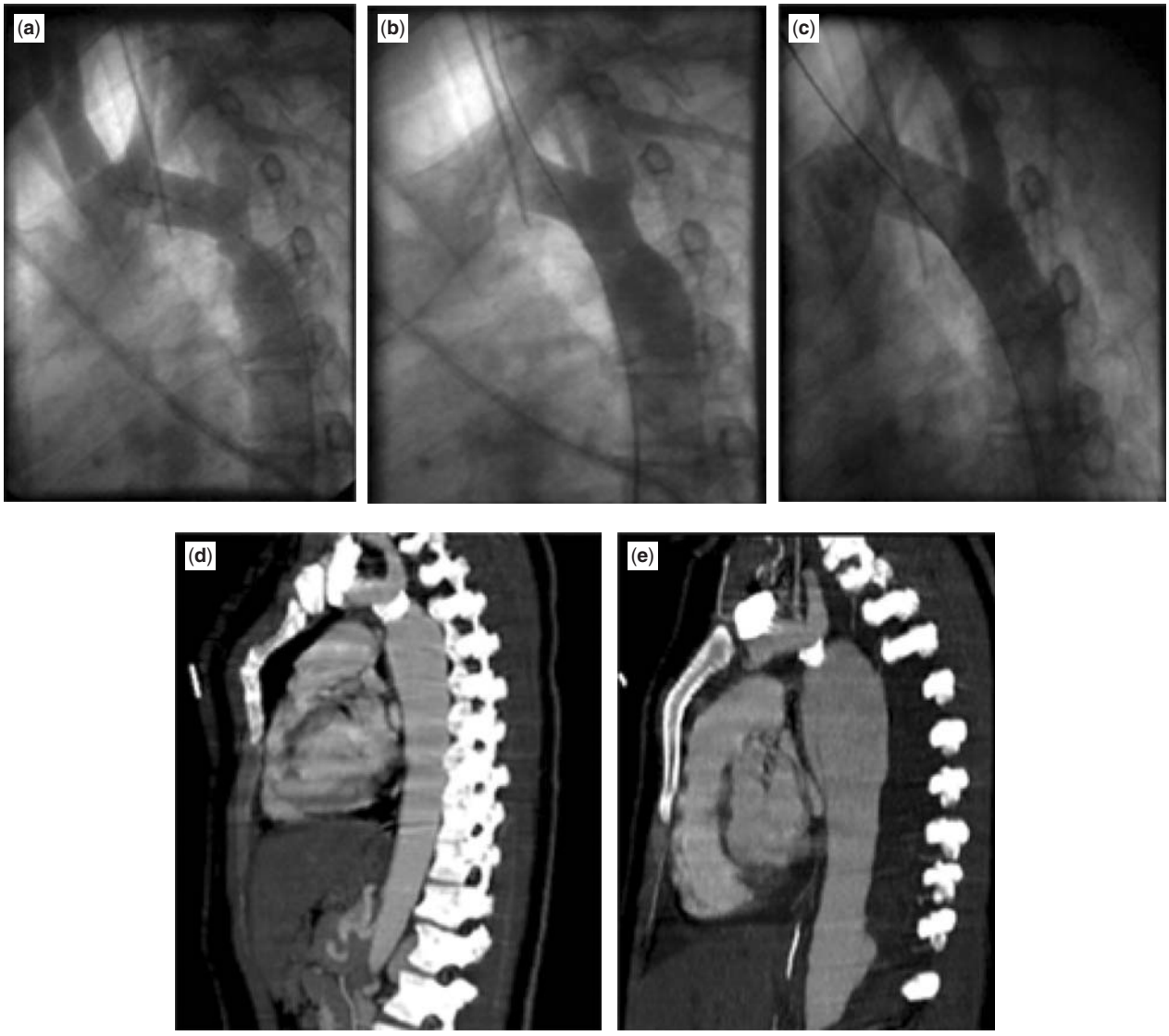

Figure 1.
*The sequence of figures show (**a**) angiography of the native coarctation; the diameter of the aortic arch, coarctation and descending aorta being 15, 6.5 and 19 millimetres respectively; (**b**) the stent initially dilated up to 14 millimetres; (**c**) the stent re-dilated up to 18 millimetres with no sign of aneurysm or dissection; (**d**) the first computed tomography scan showing dissection of the descending aorta; and (**e**) the second scan revealing the increased diameter of the dissection 3 days later.*

of elastic fibres and cystic medial necrosis. Investigation of the maternal deoxyribonucleic acid, in October 2003, showed evidence of change in the fibrillin-1 gene, as seen in Marfan's syndrome, albeit that the mother had no physicals signs of the syndrome.

Discussion

Aortic coarctation is a fairly common malformation, accounting for almost one-tenth of all congenital heart defects, which means 0.5 percent in the total population. In Turner's syndrome, the prevalence was noted at 10 percent in a series of 179 patients.[1] The management of a native coarctation of the aorta in the general population depends upon the age, morphology of the obstructive lesion, and the local institutional results for each of the four types of treatment, namely surgery, balloon angioplasty, staged dilation of a stent, or implantation of a covered stent with primary dilation to the optimal diameter. In adults, the common surgical technique is resection with end-to-end anastomosis, with a low risk of injury to nerves, chylothorax, wound infection, postcoarctectomy syndrome, and paradoxical postoperative hypertension. The most feared complications are spinal cord ischaemia, and mesenteric arteritis with infarction of the bowel. In 1984, it was already noted that surgery for aortic coarctation carries greater risks in patients with Turner's syndrome.[2]

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Balloon angioplasty was introduced over 20 years ago as the treatment for re-coarctation of the aorta, and is now considered the preferred treatment for children and adults after surgery. Potential complications of balloon angioplasty include residual aortic coarctation, aortic dissection, aortic rupture, aneurysmal formation, and femoral arterial complications. A prospective randomized study showed a higher rate of re-coarctation after balloon angioplasty, but complications observed after surgery were more serious. The difference, nonetheless, was statistically not significant.[3] Successful balloon dilation of a native aortic coarctation has been reported for patients with Turner's syndrome.[4,5] Aortic dissection after surgical repair with interposition of a graft is also reported in the setting of this syndrome.[6] Some years ago, implantation of a stent was advocated as an alternative to either surgery or balloon angioplasty.[7] More recently, a two-staged procedure has been suggested, reputedly permitting controlled injury of the aortic wall, and allowing time for healing before undertaking the second dilation half a year later.[8] Very recently, covered stents have been used to treat coarctation or re-coarctation of the aorta, especially when there has been aneurysmal formation.[9] The use of covered stents is limited, as it may not be possible to re-dilate them without damaging the covering material. To our knowledge, there are no previous reports of implantation of stents for relief of aortic coarctation in patients with Turner's syndrome. It was because of the known increase in surgical risk that we chose to implant a stent in our patient. The procedure was cautious, being planned in two stages to avoid a false aneurysm, as we were aware of the possibly more vulnerable aortic wall, knowing that the patient's mother had died from a dissection of the ascending aorta, and also aware of the association of Turner's syndrome with cystic medial necrosis of the aortic wall. At that time, a suitable covered stent for a one-stage procedure was not available in The Netherlands.

After the first stage, achieving dilation to 14 millimetres diameter, we had reduced the gradient to 15 millimetres of mercury difference in systolic blood pressure between the arm and leg. To achieve an optimal result in the second stage, we dilated the stent to 18 millimetres, and removed the gradient in pressure. This procedure shortened the stent from 15 to 11 millimetres. So, a short part of the damaged intima was not covered by the stent. It is likely that this uncovered area was the origin of the dissection of the aorta, with a possible intrinsic defect of the wall. We have no precise data, as her family did not permit an autopsy. In general, aortic dissection is a rare complication after implantation of stents for relief of aortic coarctation.[10] We need to be very cautious, however, of using the technique in patients with Turner's syndrome, or those with a family history of connective tissue disorders.

In retrospect, it would have been preferable had we accepted the fairly good result achieved after the first stage. Next time, we will implant a covered stent, dilating it to the optimal diameter, in patients with aortic coarctation and possible disorders of the connective tissues.

References

1. Gotzsche CO, Krag-Olsen B, Nielsen J, Sorensen KE, Kristensen BO. Prevalence of cardiovascular malformations and association with karyotypes in Turner's syndrome. Arch Dis Child 1994; 71: 433–436.
2. Brandt III B, Heintz SE, Rose EF, Ehrenhaft JL, Clark EB. Repair of coarctation of the aorta in children with Turner syndrome. Pediatr Cardiol 1984; 5: 175–177.
3. Hernandez-Gonzales M, Solorio S, Conde-Carmona I, et al. Intraluminal aortoplasty vs. surgical aortic resection in congenital aortic coarctation. A clinical random study in pediatric patients. Arch Med Res 2003; 34: 305–310.
4. Park Y, Sklansky MS, Shaughnessy RD, Kashani IA, Rothman A. Balloon dilatation of native aortic coarctation in two patients with Turner syndrome. Pediatr Cardiol 2000; 21: 474–476.
5. Mendelsohn AM, Lloyd TR, Crowley DC, Sandhu SK, Kocis KC, Beekman III RH. Late follow-up of balloon angioplasty in children with a native coarctation of the aorta. Am J Cardiol 1994; 74: 696–700.
6. Badmanaban B, Mole D, Sarsam M. Descending aortic dissection post coarctation repair in a patient with Turner syndrome. J Card Surg 2003; 18: 153–154.
7. Magee A, Brzezinska-Rajszys G, Qureshi S, et al. Stent implantation for aortic coarctation and recoarctation. Heart 1999; 82: 600–606.
8. Duke C, Qureshi S. Aortic coarctation and recoarctation: To stent or not to stent? J Interv Cardiol 2001; 14: 283–298.
9. Qureshi S, Zubrzycka M, Brzezinska-Rajszys G, Kosciesza A, Ksiazyk J. Use of covered Cheatham-Platinum stents in aortic coarctation and recoarctation. Cardiol Young 2004; 14: 50–54.
10. Varma C, Benson L, Butany J, McLaughlin P. Aortic dissection after stent dilatation for coarctation of the aorta: a case report and literature review. Catheter Cardiovasc Interv 2003; 59: 528–535.