Successful percutaneous stent implantation for isolated dismal transverse aortic arch kinking

Zhi-Liang Zuo, MM*, Jia-Yu Tsauo, MS, Mao Chen, MD, PhD, Yuan Feng, MD*

Abstract
Rationale: Isolated dismal transverse aortic arch kinking in adults is rare, and there is no recommended therapy at present. Percutaneous stent implantation may be an effective method to correct it and could be considered.

Patient concerns: We report a 46-year-old woman who suffered from recurrent migraine and refractory hypertension with a significant systolic blood pressure difference between upper limbs.

Diagnoses: The woman was diagnosed with isolated dismal transverse aortic arch kinking with refractory hypertension.

Interventions: Percutaneous stent implantation was performed. Due to the kinking nature of the diseased transverse aortic arch, the first covered stent moved forward to the proximal transverse aortic arch during deploying without the left common carotid artery occlusion. And then, a second stent was placed to cover the residual kinked part of the dismal transverse arch.

Outcomes: Angiography and post-procedural computed tomography angiography revealed fully corrected of the diseased segment. At 6-month follow-up after procedure, the patient was free of any symptoms and had a normal blood pressure under antihypertensive treatment.

Lessons: This case indicates that transverse aortic arch kinking in isolation can be well treated by percutaneous stent implantation in adult patients. Unlike pure aortic coarctation, elongation and bucking give the rise to the occurrence rate of stent sliding and migration and sometimes a second stent is needed.

Abbreviations: BP = blood pressure, CTA = computed tomography angiography, LCCA = left common carotid artery, LFA = left femoral artery, LSA = left subclavian artery, LVH = left ventricle hypertrophy, RFA = right femoral artery, TTE = transthoracic echocardiography.

Keywords: aortic arch kinking, percutaneous stent implantation, pseudo-coarctation, refractory hypertension

1. Introduction
Aortic arch kinking usually occurs at the isthmus segment[1,2] and the dismal transverse aortic arch kinking is rare, especially in isolation. This kind of malformation is functionally similar to the coarctation at the dismal transverse aortic arch, so patients with dismal transverse aortic kinking may present with clinical features such as hypertension.[3] The standard therapy for coarctation in dismal arch is surgical repair rather than stent implantation, which is rarely used. Currently, there is no recommended treatment for dismal transverse aortic arch kinking in adults. However, kinking is usually accompanied by elongation, which could lower the risk of the left common carotid artery (LCCA) and the left subclavian artery (LSA) occlusion. Therefore, percutaneous stent implantation could be considered. We described a 46-year-old female patient with dismal transverse aortic arch kinking who successfully underwent a percutaneous stent implantation.

2. Case presentation
A 46-year-old Chinese female patient with a history of recurrent migraine and refractory high blood pressure (BP) for >7 years was admitted to our department. The patient also had a history of neurosurgery owing to a ruptured intracranial aneurysm 7 years ago. Her BP was poorly controlled despite long-term antihypertensive treatment. Her peak systolic BP was reported at 200 mmHg, and her systolic BP of right arm fluctuated between 160 to 180 mmHg before intense antihypertensive therapy. During admitting physical examination, BP differed significantly between her left and right upper limb (143/77 and 108/70 mmHg, respectively). Her BP in the left lower limb was 117/75 mmHg and her right lower limb was 124/77 mmHg. Auscultation revealed a mild systolic murmur at the second left sternal border. Transthoracic echocardiography (TTE) showed left ventricle hypertrophy (LVH), as well as aortic arch tortuosity with local stenosis and isthmus dilation. Doppler ultrasound detected an accelerated speed of 4.3 m/s and a pressure gradient of 75 mmHg across the kinked segment of the disease aortic arch. The patient was free of any valve-related deformity or disease and the left ventricular ejection fraction was 71%. Arterial Doppler ultrasound of the upper limbs showed that the velocity of the blood
stream in the left arm had decreased, and left vertebral artery stealing was found. Computed tomography angiography (CTA) showed that the aortic arch was elongated and elevated with a tortuous kinking between the dilated LSA and the LCCA (Fig. 1A and B). In addition, the diseased aortic arch segment was also mildly calcified, and the LV was hypertrophic, which was in agreement with TTE. Furthermore, there was no significant collateral circulation. After discussion between thoracic surgeons and cardiologists, percutaneous interventional treatment was recommended to the patient, and patient consent was obtained.

The patient underwent general anesthesia in the cardiac catheterization laboratory, and a temporary pacemaker wire was positioned in the right ventricle from the right femoral vein. Both femoral arteries were punctured, and a proglide was preimplanted into the right femoral artery (RFA). A 0.035-inch guidewire was placed from the left femoral artery with a 5F catheter (VER) to the LSA for assisting location and protection from potential occlusion during stent deployment. Then, a 6F pigtail catheter was advanced to the ascending aorta from the RFA for aortography and pressure measurement. During the crossing of the diseased transverse aortic arch, the inferior kinking and stenosis could be easily corrected to some degree during the maneuvering of the pigtail catheter (Fig. 1C). Angiography presented a distorted and confined transverse aortic arch between the LCCA ostia and the LSA ostia, which was identical to the CTA images. Angiography also proved that there was dilation at the ascending aorta and the isthmus segment. Systolic pressure gradient across the bucking segment tested by the pigtail catheter was 20 mmHg. Next, a 260-cm-long Lunderquist extra stiff guidewire was inserted and the pigtail catheter was exchanged. Through this guidewire, the mid-section of a 45-mm-long covered stent graft (NuMED C-P) fitted together with a 45 x 16 mm balloon (NuMED BiB) was delivered to the kinking point with a 14F Cook sheath. Immediately after deployment, the x-ray showed that the stent had migrated forward and the upper margin of the distal diseased aortic arch was not covered (Fig. 1D). Angiography demonstrated that the LCCA ostia was mildly compressed; however, the blood flow was not influenced. Subsequently, a second 39-mm-long stent was implanted at the uncovered diseased segment (Fig. 1E). Several postdilations were performed to maximize expansion to ensure tight adhesion of the stent to the wall. During each balloon expanding, rapid pacing (180 bpm) was applied. Postprocedural angiography showed that the kinking was fully corrected, and both the LCCA and the LSA were not compromised (Fig. 1F). Pressure measurement revealed that the systolic gradient across the diseased area had disappeared, and then we withdrew the sheath and sutured the puncture sites.

After procedure, the patient was given antihypertensive medication (levamlodipine and perindopril) and the systolic pressure gradient between both upper limbs had disappeared. Postprocedural TTE demonstrated that the pressure gradient across the stents was 19 mmHg, which was significantly lower than before. Postprocedural CTA showed normal function of these stents (Figure 1G). The patient made an uneventful recovery and was discharged 1 week after procedure. One month after the procedure, the patient got a CT scan at outpatient department, and the stents did not migrate (Figure 1). At 6-month follow-up, the patient was free from any symptoms, and systolic BP of both upper limbs was between 130 and 140 mmHg.

3. Discussion

Hypertensive cardiovascular events are the most common complications in patients with aortic coarctation. For this middle-aged patient, she had a history of cerebrovascular
hemorrhage and sustained refractory hypertension with significant pressure difference between upper limbs. In combination with the TTE test, we may be reminded of transverse aortic coarctation, which is one of the most common congenital heart diseases. However, CTA demonstrated an isolated kinking at the distal transverse aortic arch, which is considerably rare. Furthermore, angiography showed that during the manipulation of the pigtail catheter across the segment of the diseased aortic arch, the kinking could be easily corrected. These informed us that it was a pseudocoarctation rather than congenital coarctation. Elongation and kinking were reported to be common in cases of pseudocoarctation. In consideration of no significant collateral arteries in CTA images, we could suspect that in the present case, the elongated transverse arch could have been present because the patient was much younger. However, it was not kinked or the kinking was not severe until recent years.

In patients with dismal transverse aortic arch coarctation, surgery repair is the standard therapy, whereas there is no recommended treatment for kinking in adults. Indeed, surgery requiring a thoracotomy has a relatively higher risk in contrast with percutaneous treatment which is minimally invasive and can be well accepted by patients. Recently, there is a systematic review reporting 20 cases of aortic arch pseudocoarctation mainly treated by surgery or managed conservatively. To our knowledge, no literature has reported percutaneous stent implantation for dismal transverse aortic arch pseudocoarctation caused by kinking in isolation. However, kinking is usually accompanied by elongation, which probably lowers the risk of LCCA and LSA occlusion during stent deployment. Therefore, in present case, transcatheter stent implantation could be taken into consideration. During the deployment of the first stent, the stent partially migrated to the proximal segment of the transverse aortic arch. The cause of this sudden migration of the stent after deployment could be because of the kinking nature of the diseased aortic arch rather than coarctation. After the procedure, the pseudocoarctation was erased. And then the pressure gradient between the upper extremities had disappeared and her hypertension had become easier to be controlled. In Surlan et al.'s report, the self-expanding Gianturco-Rosch Z-stents were used to exclude the thoracic aortic aneurysm and then led to proximal stent prolapse into the aneurysm in the long-term follow-up. In our experience, balloon-expanded stents are less likely to migrate in contrast with self-expanding stents. However, longer follow-up and careful future observation for this patient are necessary, as restenosis and hypertension could affect the outcome of survival.

4. Conclusion

In conclusion, isolated dismal transverse aortic arch kinking is uncommon and can be well treated with percutaneous stent implantation. Unlike pure aortic coarctation, elongation and bucking give the rise to the occurrence rate of stent sliding and migration and sometimes a second stent is needed.

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