

Short- and mid-term effects of covered stent implantation on extremity findings and heart failure in Parkes Weber syndrome: a case report

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Background

Parkes Weber syndrome (PWS) is a congenital disease characterized by vascular malformations, such as arteriovenous fistulas (AVFs). It frequently presents with overgrowth of a lower limb and high-output heart failure. The main treatment is to close vascular malformations. Surgical excision or endovascular coil insertion was performed in a few patients with AVFs. However, vascular covered stent implantation has not been used for treating PWS.

Case summary

A 15-year-old male patient with PWS presented to our hospital because of dyspnoea and massive left upper limb swelling. After initial examination and left upper limb angiography, his symptoms and findings were attributed to the presence of high-flow large AVFs despite the presence of many coils previously inserted. We decided to implant a covered stent along the AVFs between the subclavian and axillary arteries. After stent implantation, the patient's complaints and findings improved during the early term but they relapsed at the 6th month after percutaneous intervention.

Discussion

Here, we report for the first time the use of covered stent implantation and its short and 6 months results in a patient with PWS. Although initial improvements were noted, the clinical outcome at 6 months after stent implantation was poor. This was probably associated with the presence of widespread subtle AVFs or collateral connections among the existing AVFs. Based on our result, we propose that closure of large AVFs is not useful and more definitive interventions, such as limb amputation may be required earlier.

Keywords

Parkes Weber syndrome • Arteriovenous malformation • Covered stent implantation • Case report • Heart failure

Learning points

- There may be many subtle arteriovenous fistulas (AVFs) other than existing AVFs along the affected limb in Parkes Weber syndrome (PWS).
- Closure of the obvious AVFs by vascular instruments can lead to the activation of subtle inactive ones.
- More definitive treatment approaches, such as amputation of involved limb may be thought in early term of disease in patients with PWS who have large high-flow AVFs.

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Introduction

Parkes Weber syndrome (PWS) is a rare congenital disorder associated with severe vascular malformations, such as arteriovenous fistulas (AVFs).¹ Arteriovenous fistulas can cause limb swelling and high-output heart failure associated with impaired life quality and decreased life span.² Although surgical resection and percutaneous coil embolization have been used to remove vascular malformations, no data are available on covered stent implantation. Moreover, very few studies have reported mid- and long-term results of surgical and endovascular techniques for treating vascular malformations in PWS. Therefore, there is no definitive treatment algorithm and patient management is mostly tailored according to their individual characteristics. In this case report, we describe for the first time, short- and long-term (at 6th month) results of a patient with PWS who had high-output heart failure and left upper limb swelling because of AVF in the subclavian artery region and was treated by covered stent implantation.

Timeline

2010	Symptoms of heart failure started
February 2019	Patient presented to our clinic. Initial limb extremity angiography and covered stent implantation were made.
March 2019	Patient's complaints and findings progressively improved. Heart failure medication reduced.
April 2019	Second angiography was made and follow-up with medical treatment was decided.
July 2019	Patient's limb swelling and dyspnoea started again.

Case presentation

A 15-year-old male patient with PWS presented with complaints of shortness of breath and massive left upper limb swelling. Clinical examination revealed that he had dyspnoea at both rest and exercise despite taking heart failure medications including digoxin, metoprolol, furosemide, spironolactone, and enalapril. He also had a port-wine stain on his left pectoral area, severe pain and massive swelling of the left upper extremity, and finger cyanosis on the left hand (*Figure 1*). His blood pressure was 140/60 mmHg and pulse was 120 b.p.m. There were moderate peripheral oedema and jugular venous distension. Systolic flow murmurs and third heart sound were heard by cardiac auscultation. Transthoracic echocardiography revealed normal cardiac chamber dimensions and hyperdynamic cardiac function which reflected the presence of high-output heart failure. He had previously undergone vascular coil insertion to occlude AVFs in another centre (*Figure 2*). We performed left upper extremity angiography to determine whether the AVFs still existed because the patient's complaints persisted despite previous coil insertion. In angiography, we realized that patient's high-flow AVFs were still active



Figure 1 Massive oedema and cyanosis on the left upper extremity at presentation.

([Supplementary material online, Video S1](#)). After a detailed evaluation of brain circulation, two covered stents (Atrium Advanta V12 Balloon Expandable Covered Stent, 9 mm × 59 mm × 120 cm and 8 mm × 59 mm × 120 cm) were sequentially implanted along the AVF region between the proximal part of the subclavian artery and distal part of the axillary artery (*Figure 3*). After stent implantation, smaller new AVFs were developed both at distal part of stents (along the axillary artery) and near the elbow ([Supplementary material online, Video S2](#)). We thought to occlude them by using a flexible covered stent implantation subsequently. After this initial covered stent implantation, left upper limb swelling and finger cyanosis progressively disappeared (*Figure 4*). Furthermore, patient's dyspnoea and his port-wine stain progressively improved. The intake of heart failure medications decreased (digoxin was discontinued; metoprolol dose was reduced; and furosemide dose was initially discontinued but later he needed reduced dose drug) during this early period. After 2 months, we attempted to implant a flexible covered stent to axillary AVF. Unfortunately, during control angiography, we noted overgrowth of previous small axillary AVFs and the development of many new fistulas along the arm ([Supplementary material online, Video S3](#)). Thus, we abandoned the idea of additional stent implantation and decided to follow the patient clinically. Unfortunately, this early-term improvements were not permanent and his clinical complaints and findings recurred at 6 months. He had to fully restart his heart failure medication regimen including metoprolol due to increased and uncontrolled heart rate. In addition, clinical examination of the left upper extremity revealed massive extremity swelling, large ulcers

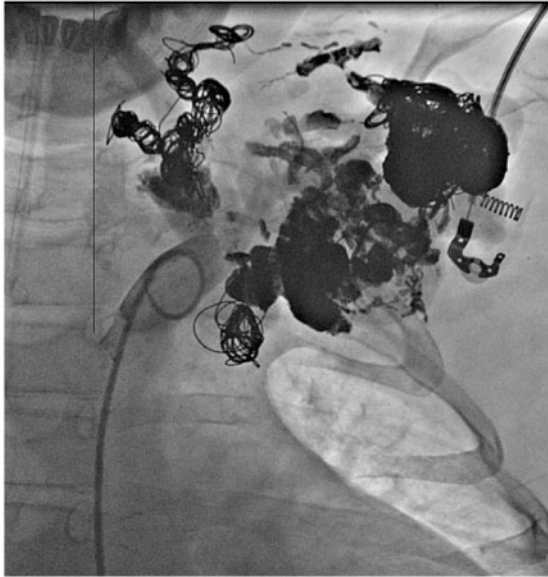


Figure 2 Many coils were previously inserted at another centre for treating arteriovenous fistulas.

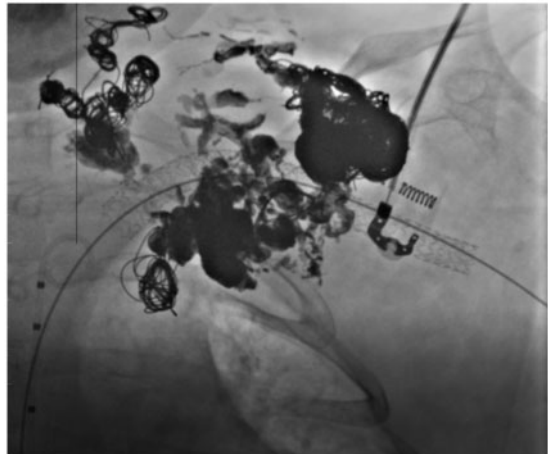


Figure 3 Two vascular covered stents were successfully implanted along the arteriovenous fistula region between the subclavian artery and the axillary artery.

on the forearm, and finger cyanosis except for port-wine stain (Figure 5).

Discussion

Parkes Weber syndrome is a rare congenital disorder of the vascular system associated with vascular malformations such as high-flow AVFs and limb hypertrophy.¹ Clinically, PWS can cause limb swelling and high-output heart failure that limits both quality of life and life



Figure 4 Left upper limb findings improved during early term after stent implantation.

span. Moreover, limb amputation might be required in some cases. One of the main treatment purposes is to control vascular malformations. Surgical and percutaneous coil embolization have previously been used for this purpose. All these treatment modalities can lead to initial clinical improvement. However, to date, only one follow-up study on long-term outcome of these techniques has been reported in literature. Thus, mid- and long-term effects of invasive treatment for such malformations are unknown. In the present case, we report for the first time both the use of percutaneous covered stent implantation for vascular malformations and its initial and 6th-month follow-up data in a patient with PWS. After stent implantation, heart failure and limb swelling improved during the early months. However, during follow-up, the patient re-experienced his initial symptoms almost at the 6th month after implantation.

We thought that these findings were due to maturation of newly developed small AVFs along the distal portions of the left arm. As an explanatory mechanism for this maturation, we proposed that patients with PWS probably have widespread inactive fistulas in close and distant areas of existing AVFs along the involved limb. When existing AVFs are occluded by surgical or percutaneously, subtle inactive fistulas become active. As another explanation, we assume that there are diffuse collateral connections among the widespread AVFs. Thus, high-flow AVFs are still maintained although they are closed.

There is no yet definitive treatment for vascular malformations. There are three basic targets in the management of this disease: to



Figure 5 Massive left upper extremity swelling, large ulcer on the forearm, and finger cyanosis developed again at the 6th month after stent implantation.

treat existing vascular malformations, to prevent the development of new ones, and to improve the quality of life. Presently, the management of PWS is focused mainly on increasing the quality of life by excision or occlusion of existing vascular malformations. Unfortunately, extremity amputation is required to improve the quality of life in some cases such as those with large high-flow AVFs. Currently, antiangiogenic drugs, such as bevacizumab and ranibizumab are used to decrease angiogenesis in some cancer types, including colorectal and neovascular age-related macular degeneration. Although these drugs have many cardiovascular adverse effects, such as heart failure, QT prolongation, and arterial and venous thrombus, they may be used to manage PWS in the future.³ Therefore, further studies are needed to investigate the possible role of antiangiogenic agents in patients with PWS.

The main differences between the present case and previously reported cases can be summarized as follows: first, the affected limb was mostly lower extremity in previous PWS but the upper limb was involved in the present case^{2,4}; second, to our knowledge, this is the first case of the use of covered stent; and third, a previous case reported only early-term result after endovascular coil insertion for treating right upper limb AVFs, whereas we reported both short-term and 6th month after implantation results.⁵ Therefore, we think that this case report offers additional data on such cases to literature.

Conclusion

On the basis of our case findings, we think that the closure of large AVFs was not useful and more definitive interventions, such as limb amputation may be useful at earlier stages in patients with PWS who have severe clinical complications, such as massive limb swelling and heart failure. Alternatively, antiangiogenic drugs may offer an option for the treatment of this disease in the future, but further studies are needed.

Lead author biography



Dr Zeydin Acar received his MD from Ondokuz Mayıs University, Samsun, Turkey. He also completed his cardiology education at Ondokuz Mayıs University. Since 2013, Dr Acar is currently working at Medical Park Karadeniz Hospital, Trabzon, Turkey. He obtained Assistant Professor Degree from Altınbaş University from Istanbul at 2015. Interventional cardiology is one of his main research areas. He has published many scientific papers in respectable journals of 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 author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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