This unusual arteriopathy was first reported in 1908 by the Japanese Ophthalmologist Takayasu. He described a young woman with cataracts and unusual wreath-like arteriovenous anastomoses surrounding the optic papillae. The optical findings were also found in patients with absent radial pulses and later in patients with other occlusions of the aorta and its branches. The disease has also been termed “pulseless disease”, “aortic arch syndrome”, “reversed coarctation”, “occlusive thromboarteriopathy” and “young female arteritis”. No specific cause has been identified and some of the difficulties that may arise in management are well illustrated by the following case.

Case Report. A 31-year-old female presented with two episodes of syncope during mild exertion in the previous month. She was an unmarried Thai student in Australia, a non-smoker who drank little alcohol and formerly played competitive basketball. A complete medical assessment three years before revealed no abnormalities. There was a family history of hypertension. One month prior to admission, she had a syncopal episode while walking. Initially, she was thought to have mitral regurgitation and was started on heart failure medication. A second syncopal episode with seizure activity occurred one week later. She described a four-year history of dizzy turns on exercise, occasional heaviness and tingling in her right arm and mild exertional dyspnea. There was no history of claudication, Raynaud’s phenomena, fevers, myalgia, rash or weight loss. Examination revealed a regular pulse, but all peripheral pulses were difficult to palpate. There was a prominent thrill at the base of the neck on the right side. The blood pressure was 90/60 mmHg measured in the right arm, 60/40 mmHg in the left arm, and 100/60 mmHg in the right leg. The heart sounds were normal and there was a loud systolic murmur throughout the precordium, neck and back. A loud abdominal bruit was present and the neurological examination was normal. A provisional diagnosis of Takayasu’s Arteritis was made. Investigations included an ECG showing sinus rhythm with voltage evidence of left ventricular hypertrophy and a normal chest x-ray. The echocardiogram showed no chamber enlargement, normal ventricular systolic function, mild mitral regurgitation, and high velocity flow at the base of the right side of the neck. Carotid duplex revealed > 80% stenosis in the right common carotid artery extending from its origin for 4 cm, a patent right vertebral artery, > 80% long left common carotid artery stenosis and a patent left vertebral artery with retrograde flow. The patient was admitted for 4-vessel angiography, which showed multiple large-vessel stenoses, including the distal thoracic aorta (Figures 1 and 2). There was a systolic gradient of 100 mmHg across the aortic stenosis with the proximal aortic pressure being 215/77 mmHg. The CRP was 35 (0–8 mg/L), the ESR > 100 (0–7 mm/hour), Interleukin 6 (20) and there was a mild normochronic, normocytic anemia (Hb, 110%). The von Willebrand factor was normal at 115 (50–200), the ANA, ENA and ANCA were negative and there was a polyclonal hypergammaglobulinemia. The heart failure medication was ceased and prednisone 50 mg and aspirin 150 mg daily commenced. She was discharged. One week later, the ESR was 54 and CRP was 4; at two weeks she was asymptomatic. A further syncopal episode on exertion occurred four weeks later and at that time the ESR and CRP were normal. The plan was to repeat the angiography at three months with a view to intervention.

Discussion. The presentation, clinical findings and investigations are typical of Takayasu’s Arteritis. Prednisone appeared to reduce the inflammatory response and repeat angiography is planned in three months to assess the lesions. The hypertension is
presumably due to the acquired aortic coarctation. Its management is complicated by the risk to the cerebral circulation if the blood pressure is reduced and by the difficulties in monitoring the blood pressure non-invasively. Successful percutaneous angioplasty with or without stenting has been reported and may be undertaken in this patient after angiographic review at three months. The initial strategy would probably be a serial approach to the lesions with the left subclavian artery and the aortic coarctation. Hopefully, relief of the left subclavian stenosis would improve the cerebral circulation and reduce the risk of a subsequent procedure to the right innominate, common carotid and subclavian arteries if required. Correction of the aortic coarctation may resolve or reduce the hypertension. A serial surgical approach would also be feasible and successful surgical results have been reported. The prognosis in this syndrome is variable and depends on the response to medication, the presence or development of complications and the result of intervention. In this patient, the risk of interventions is significant in view of the severity and the site of the lesions. In summary, this 31-year-old Thai female with Takayasu’s Arteritis with multiple stenoses affecting the aortic arch branches and the thoracic aorta has a high risk of stroke and is a challenge in management.

REFERENCES