

Hypertension in the Young Adult – Come Feel the Pulse

L F Hsu, K N Sin Fai Lam, C Rajasoorya and L S Chew

SUMMARY

Hypertension occurring in teenagers and young adults is uncommon. Though the most common form is still essential hypertension, secondary causes are more commonly found here than in older adults. Renal, cardiovascular and endocrine diseases constitute most of these causes. Coarctation of the aorta is the most common cardiovascular cause of hypertension, and its importance lies in the fact that it is correctable, and that its persistence often leads to dangerous complications and early death. The cardinal sign of differential pulse and blood pressures between the upper and lower limbs can be detected clinically. Hence, the importance of a detailed physical examination in all young hypertensives, including palpation of all the pulses, cannot be overemphasized.

We present 2 hypertensive young men who were found to have isolated coarctation of the aorta. The lesion in the first patient was located postductally just distal to the left subclavian artery. This area has been found to be the most common site of coarctation. The second patient had an unusual mid-thoracic coarctation. The clinical and radiological features as well as complications are highlighted. In young hypertensive patients, a high index of suspicion may enable the physician to make a timely diagnosis and hence avert the potentially disastrous complications that may arise in undetected cases.

Key words: coarctation, aorta, hypertension, young, aortography

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INTRODUCTION

Hypertension occurring in children, adolescents and young adults is uncommon and almost always sparks off an intense search for an underlying aetiology. Though essential hypertension is still the commonest form here, secondary hypertension occurs with greater frequency than in adults and is most often due to renal disease,

followed by cardiovascular and then endocrine disorders⁽¹⁾.

Coarctation of the aorta is the most common cardiovascular cause of hypertension. It is also the fourth most frequent form of symptomatic congenital heart disease, constituting between 6% to 9% of congenital heart defects in children^(2,3) and about 15% in adults⁽²⁾. In its most basic form, it consists of haemodynamically significant narrowing of a segment of the aorta. This is usually located proximal to the insertion of the ductus arteriosus (preductal) in infants, while in older children and adults, it is located at the level of the ductus (juxtaductal) or distal to it (postductal). The majority occur just distal to the ductal insertion^(3,4). We present 2 patients with isolated aortic coarctation, the first in the usual and the second in an unusual site.

CASE REPORTS

Case 1

A 20-year-old Chinese soldier was referred for assessment of hypertension detected during routine medical screening. He complained of a month-long history of headache, retrosternal discomfort and inappropriate lower limb fatigue on exertion. His blood pressure was found to be 170/100 mmHg in both arms, his femoral pulses were barely palpable, and there was clinically evident radio-femoral delay. No praecordial or posterior interscapular bruits were detected.

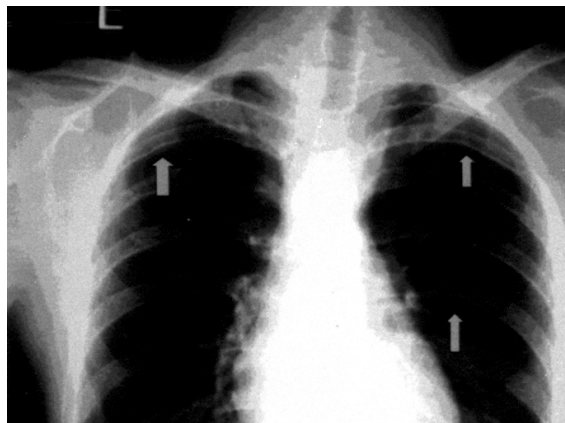


Fig. 1 Chest radiograph of Case 1 showing typical notching of the undersurfaces of the ribs posteriorly (arrows).

Department of
Medicine
Alexandra Hospital
378 Alexandra
Road
Singapore 159964

L F Hsu, MBBS,
MRCP (UK)
Medical Officer
(Specialist)

K N Sin Fai Lam,
MBBCh, FRCPI,
FCCP
Senior Consultant

C Rajasoorya,
MBBS, FRCP
(Edin), FAMS,
FACE
Consultant

L S Chew, MBBS,
PPA
Clinical Associate
Professor and
Senior Consultant

Correspondence to:
Dr K N Sin Fai Lam
Tel: 470 8353
Fax: 479 3183



Fig. 2a Aortogram of Case 1. The catheter has passed through the coarctation, and injection of contrast reveals the site of obstruction to be just distal to the left subclavian artery.



Fig. 2b Aortogram of Case 1. A later phase of the aortogram highlighting the collateral circulation.

The electrocardiogram showed left ventricular hypertrophy and a chest radiograph revealed notching of the inferior borders of his ribs bilaterally (Fig. 1). An aortogram showed a tight stenosis of the thoracic aorta just distal to the left subclavian artery (Fig. 2a) with very large collateral vessels (Fig. 2b). Spiral

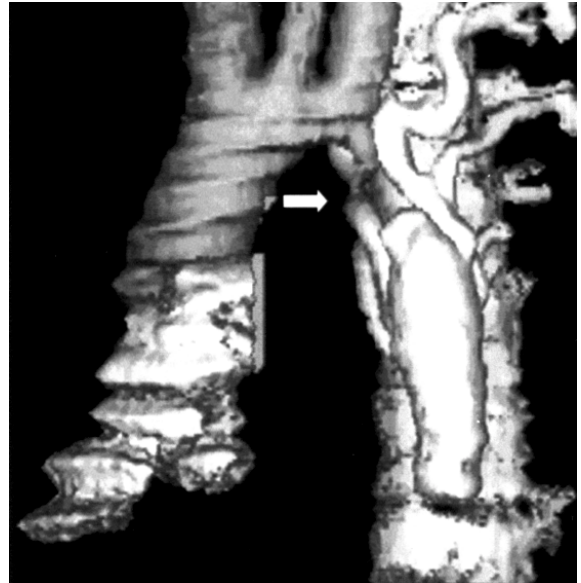


Fig. 3 Spiral CT scan with 3-dimensional reconstruction of the aorta, showing the site of the lesion and the extensive collateral circulation.

computed tomography (CT) scan with 3-dimensional reconstruction of the images (Fig. 3) revealed coarctation of a short segment of the aorta immediately after the origin of the left subclavian artery, with pre and post-stenotic dilatation. Very large collateral vessels were also visualised.

At operation, the lesion was found at the isthmus just distal to the left subclavian artery and ligamentum arteriosus, with multiple large dilated intercostal arteries. Excision and repair with a Dacron graft was performed, and the patient recovered uneventfully post operatively. He has since remained normotensive.

Case 2

A 15-year-old schoolboy was referred for assessment of hypertension detected on routine medical screening at school. He was asymptomatic. Clinical examination revealed a blood pressure of 180/90 mmHg with no differential pressure between both arms. He had weak femoral pulses with radio-femoral delay, a systolic murmur over the praecordium and an audible posterior interscapular bruit.

An electrocardiogram showed left ventricular hypertrophy and his chest radiograph showed bilateral rib notching. Aortography confirmed a severe coarctation at the mid thoracic level (Figs. 4a, b, c) with large collateral vessels (Fig. 5). Echocardiography did not reveal any associated cardiac defects. The patient refused surgery despite full knowledge of the grave prognosis and possible complications.

DISCUSSION

Coarctation of the aorta is a congenital anomaly of uncertain aetiology which is widely regarded as a



Fig. 4a Aortogram of Case 2. The site of coarctation is in the mid-thoracic aorta.



Fig. 4b Aortogram of Case 2. Due to the severe narrowing, the catheter was unable to negotiate the coarctation initially.

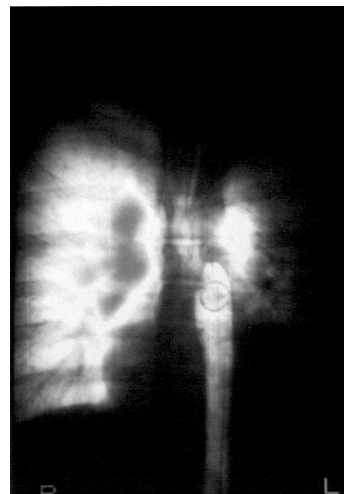


Fig. 4c Aortogram of Case 2. Another view showing the level of the lesion.



Fig. 5 A later phase of the aortogram of Case 2, showing the numerous collateral vessels.

'treatable' cause of hypertension. It predominates in males, with a sex incidence of as high as 3:1, and has been reported in families⁽⁴⁾. It is associated with Turner's syndrome in about 20% of affected females⁽⁵⁾.

In older children and adults, the underlying abnormality consists of a localised diaphragm-like infolding of the aortic media, forming a ridge which projects into the lumen, causing eccentric narrowing. The ridge almost invariably contains ductal tissue, which also extends around the aorta in a circumferential sling^(4,6). This characteristic histological finding has led to the postulation that the development of the coarctation is somehow related to the closure of the ductus arteriosus⁽⁶⁾. In older children, the majority of lesions are found distal to the ductal insertion and the obstruction commonly worsens with age⁽²⁾. This led to

the conclusion that all coarctations develop in a preductal position and migrate to a postductal position with age, while intimal proliferation further narrows the lumen and fibrosis and degenerative changes may occur⁽⁶⁾. However, this does not satisfactorily explain the occurrence of coarctation in more distal sites, both in the thoracic aorta, as in Case 2, and the abdominal aorta. In these cases, both embryological as well as inflammatory mechanisms have been proposed, although the actual aetiology remains uncertain⁽⁷⁾.

Irregardless of the site of narrowing, blood flow around the obstruction is accomplished by the development of a collateral circulation through the spinal, intercostal, internal mammary, epigastric and lateral thoracic vessels, as well as the periscapular network⁽⁵⁾, and the mediastinal collateral vessels⁽⁸⁾. These vessels tend to become more dilated with age and may erode the inferior borders of the ribs, producing the classic rib notching as seen on the chest radiographs of our patients.

Symptoms are usually found during 2 stages of life. About 10% of infants develop heart failure during the early weeks and months of life⁽³⁾. Most patients, however, have few symptoms in the first 3 decades of life. Thereafter, they eventually develop symptoms related to hypertension and its complications, while some may complain of fatigability of the lower limbs during exercise, as in Case 1. The cardinal physical sign of aortic coarctation is differential blood pressure and pulses between the upper and lower extremities. The leg recording should be at least 20 mmHg lower than the arm⁽⁹⁾.

Occasionally the patient may have hypertrophy of the upper extremities with underdeveloped lower limbs⁽⁴⁾, while superficial collateral vessels may be palpated over the scapular region. Cardiac findings may

include a thrill in the suprasternal notch, left ventricular hypertrophy and systolic and diastolic murmurs which can originate from the area of coarctation, the collateral arterial flow or associated valvular defects or shunts⁽⁴⁾.

Other than the classical rib notching, especially prominent on the third through the seventh ribs, the chest radiograph may also show a 'figure-of-three' sign, which is caused by the combination of a dilated left subclavian artery above the coarctation and the post stenotic dilation of the descending aorta below⁽⁴⁾.

Though aortography remains the standard test for diagnosis and assessment, non-invasive methods have become increasingly utilised due to the wider availability of sophisticated technology. Much of the data previously obtained by cardiac catheterisation and aortography can now be obtained with good accuracy by 2-dimensional echocardiography with Doppler studies, magnetic resonance imaging⁽⁹⁾, and spiral CT scan with image reconstruction⁽¹⁰⁾, and example of which was shown in Case 1 (Fig. 3). However, aortography and cardiac catheterisation is still important in delineating the precise anatomy of the lesion and aortic arch vessels, the presence and extent of collaterals, and the presence and significance of associated lesions and complications⁽³⁾.

Most patients with coarctation undetected during infancy have few related difficulties in the first 3 decades of life. Thereafter, the incidence of complications and risk of mortality increase if untreated. The mean survival of unoperated patients is 35 years, and survival past 50 years is unusual⁽³⁾. Heart failure is present in two thirds of patients above 40 years and is often related to associated valvular, hypertensive and coronary heart disease. Strokes occur in approximately 6% and its incidence increases with the chronicity of hypertension. Infective endocarditis was reported in 2% while 3% had aortic dissection or rupture⁽³⁾.

With these data, it becomes evident that the timing of correction is of utmost importance, with the best results obtained at 3 to 5 years of age. Correction during infancy is associated with a higher incidence of residual or recurrent coarctation, while later correction is associated with residual hypertension in 25-50% of patients⁽³⁾. Survival rates also decline the later the procedure is performed in life, with the shortest survivals in patients over the age of 40 years⁽²⁾.

Corrective surgery is the definitive treatment for aortic coarctation, though it is associated with several important complications. These include 0.4% incidence of lower limb paralysis due to spinal cord ischaemia intra-operatively⁽²⁾, and post-operative complications such as bleeding, paradoxical hypertension, infective endocarditis, chylothorax, laryngeal palsy, stroke, aortic dissection and graft aneurysm⁽³⁾. Transluminal balloon

angioplasty, which has had excellent results in the treatment of post-operative restenosis, has also been shown to be as effective as surgery in native coarctation^(11,12). Restenosis and aneurysms appeared to be more frequent with balloon angioplasty, while neurological complications were more frequently seen with surgery⁽¹¹⁾.

CONCLUSION

Coarctation of the aorta is an important cause of hypertension in young adults. Fortunately a clinical diagnosis can often be made easily based on the classical findings of differential pulses and blood pressures between the upper and lower limbs, and the radiographic features. This condition should be actively considered in all hypertensive young patients as it is eminently correctable and a delayed or missed diagnosis may result in potentially serious consequences for the patient.

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