SYSTEMATIC ANALYSIS OF THE DOPPLER TRACINGS

CASE 1

In both tracings it appears as if the patient is in a regular atrial rhythm on evaluation of the ECG recordings. In the CW Doppler tracing the cursor is well aligned with the descending portion of the aortic arch (Figure 7).

Evaluation of the flow pattern reveals high velocities during systole (peak systolic gradient obtained with the simplified Bernoulli equation 42mmHg) with a persistent gradient during diastole down the arch (a "diastolic tail"). In the PW Doppler tracing the sample volume is placed adequately in the descending aorta. Evaluation of this flow pattern reveals continuous forward flow throughout the cardiac cycle and an absence of normal pulsatile flow (Figure 8).

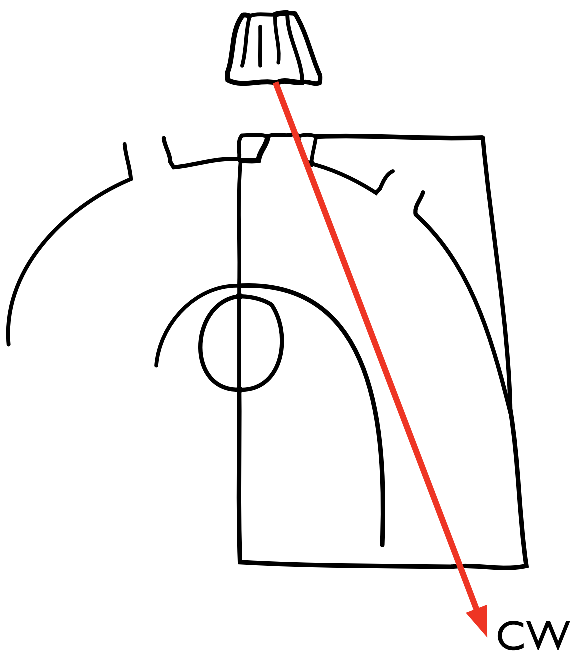

FIGURE 7: Suprasternal long axis aortic arch view showing correct alignment of cursor for CW Doppler down descending thoracic aorta.

Further discussion of Case 1

This patient was referred for an evaluation for CoA. Echocardiography is often used in the initial assessment of patients with CoA and in their follow-up after intervention. Evaluation with two-dimensional and colour Doppler echocardiographic techniques can be difficult especially in adults who have a limited suprasternal window. Analysis of CW Doppler across the CoA site and PW Doppler at the abdominal aorta are used for the indirect evaluation of CoA.[1] As with most stenotic lesions, blood accelerates through the narrowing in the aorta giving rise to the high velocities observed in systole. Dilatation of the precoarctation aorta in systole results in a persistence of stored upstream energy (depending on lesion severity and compliance of the aorta) and this stored energy is released downstream in diastole and the precoarctation walls contract leading to the diastolic tail observed in CoA.[2]

It is important to note that pressure gradients alone as an index of aortic narrowing are often inadequate because Doppler velocities are affected by cardiac output, lesion length, the presence of collateral networks and aortic compliance.[1] For instance, a patient with a severe coarctation can have a low trans-coarctation gradient if they are very well collateralised. For this reason a low gradient does not exclude severe coarctation.

The simplified Bernoulli equation (Gradient = $4V^2$) may be used if the velocity before the coarctation is known to be less than 1m/s. This is often not the case and if the velocity before the coarctation is more than 1m/s the modified Bernoulli equation (Gradient = $4\{V_2 - V_1\}^2$), which includes pre-obstruction velocity (V_1) in pressure gradient calculation, should rather be used. Moreover, abnormal arterial stiffness invalidates certain assumptions of the modified Bernoulli equation and the gradient is most likely overestimated when measured by flow acceleration. Loss of normal pulsatile flow and continuous anterograde diastolic flow in the abdominal aorta ("diastolic run-off") is a reliable indicator of

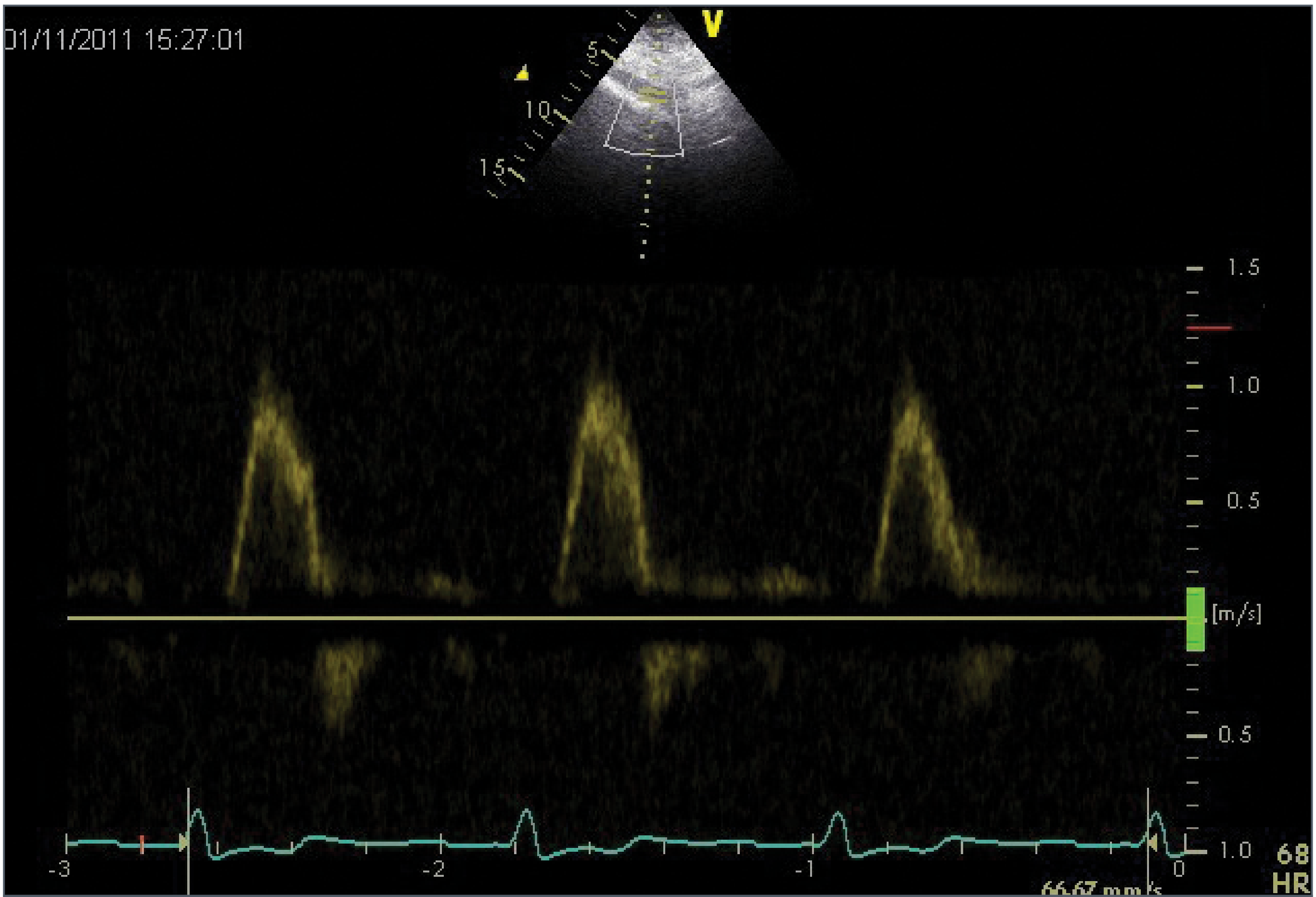

FIGURE 8: PW Doppler in the abdominal aorta (acquired via subcostal window) from a normal patient without coarctation demonstrating normal triphasic pulsatile flow. This flow pattern excludes the presence of significant coarctation.

significant CoA.[3] These Doppler tracings confirm the presence of a significant CoA, irrespective of the trans-coarctation gradient. In this patient, the correct answer is (b).

In severe aortic regurgitation (AR) holodiastolic flow reversal occurs because a column of blood flows up the aorta throughout diastole into the left ventricle through an incompetent aortic valve. Answer (a) is therefore incorrect. Even if severe AR and CoA were to coexist within the same patient, one would still expect to see a degree of flow reversal up the arch. Answer (c) is therefore also incorrect. Answer (d) is incorrect and will be discussed in more detail in the following cases.

CASE 2

This patient also appears to be in a regular atrial rhythm by analysis of the simultaneous ECG recordings. The cursor is well aligned with the descending part of the arch in the CW tracing. The systolic velocities down the arch are high as in the first case discussed (peak systolic gradient 60mmHg by the simplified Bernoulli equation), but unlike case 1, there now appears to be a persistent gradient during diastole in the opposite direction; holodiastolic flow reversal is present. This is confirmed on the PW Doppler tracing obtained with a sample volume in the proximal descending arch.

Further discussion of case 2

This patient is known to have had a CoA repair so interrupted aortic arch – answer (d) is incorrect. He also has a bicuspid aortic valve which is associated with AR, either due to the valve being abnormal itself or to root dilatation secondary to the associated aortopathy. The presence of holodiastolic flow reversal in the aortic arch is a sensitive (albeit less specific) sign of severe AR on echocardiography and therefore this patient has significant AR which makes answer (b) unlikely. Importantly, the presence of significant AR and the associated aortic flow reversal modifies the Doppler tracings expected in significant recoarctation. The presence of severe AR will make the persistent diastolic tail of significant recoarctation disappear because the AR causes flow in the aorta to reverse and therefore persistent forward flow in diastole cannot occur. However, the velocity in the arch obtained by PW Doppler is 2m/s and this velocity jumps to almost 4m/s obtained by CW Doppler down the descending thoracic aorta, indicating a narrowing in this portion of the aorta. This jump in flow velocities cannot be attributed to severe AR, and this makes answer (a) unlikely. The presence of a jump in velocities in the arch as well as holodiastolic flow reversal in this patient is compatible with severe aortic regurgitation in the presence of some degree of recoarctation. The correct answer is therefore (d) and further non-invasive testing either by CT but preferably MRI is recommended to adequately evaluate the degree of recoarctation.

CASE 3

This patient also appears to be in a regular atrial rhythm. The cursor is well aligned down the descending part of the aortic arch in the CW Doppler tracing, but in this case there is no appreciable systolic gradient and no diastolic tail. The PW Doppler tracing, with the sample volume placed accurately in the abdominal aorta, reveals normal pulsatile flow.

FIGURE 9: CT demonstrating a 2.5mm interruption of the aortic arch just distal to the left subclavian artery. This suggests IAA Type A.

Further discussion of case 3

A story of a young hypertensive with prominent radio-femoral delay is very suggestive of CoA. Surprisingly in this patient, no significant gradient is seen in the arch, there is no diastolic tail and the abdominal aortic Doppler shows pulsatile flow which strongly argues against coarctation. The differential for this scenario would include interruption of the aortic arch (IAA) and "functional" interruption of the aorta by a critical coarctation where forward flow is almost absent and extensive collateralisation has occurred. Because the aorta is interrupted and not stenosed, no flow acceleration occurs at the site of obstruction and therefore no systolic gradient or diastolic tail are observed. Normal pulsatile flow may be observed in the abdominal aorta due to the extensive collateralisation. IAA is defined as a discontinuity in the aortic arch and is extremely rare in adults, with only 34 cases described in the literature to date. IAA is divided into 3 types: A – discontinuity distal to the left subclavian artery (30 - 40%); B – discontinuity between the left subclavian and the left carotid arteries (55 - 60%); and C – the most uncommon type - interruption proximal to the left common carotid artery.[4]

Distinguishing between severe CoA and Type A IAA can be very difficult. There are helpful images on CT and MRI which may facilitate the differentiation,[4,5] but caution should be exercised in diagnosing IAA with these imaging modalities alone due to the effect of partial volume rendering the narrow neck of a severe coarctation invisible. Transoesophageal echocardiography can also help to demonstrate colour flow from the descending thoracic aorta to the abdominal aorta distal to a severe CoA.[6] In the case of IAA it is practically impossible to pass a catheter from the femoral artery to the proximal aorta, so if suspected, right radial angiography should preferably be performed if catheterisation is planned.

In this patient, a significant CoA would be expected to give the Doppler signs of CoA as discussed above, so answer (b) is incorrect. The absence of holodiastolic flow reversal excludes both diagnosis (a) and (c) (as explained in cases 1 and 2). This patient did indeed have IAA Type A (Figure 9). The correct answer is (d).

LESSONS LEARNED AND CLINICAL IMPLICATIONS

Patients with significant CoA and recoarctation have distinguishing features in the descending thoracic and abdominal aortic flow velocities and profiles, such as the presence of a diastolic tail and continuous abdominal aortic flow. These Doppler profiles become unreliable in the setting of associated AR and alternative forms of non-invasive imaging should be used to evaluate for CoA. If CoA is strongly suspected clinically and the Doppler profiles are normal or near normal, the extremely rare case of IAA should be suspected.

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