Transcranial Doppler in the blue stander: the platypnoea-orthodeoxia syndrome revisited

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DESCRIPTION

A young man was referred to our neurosonology laboratory after an endovascular closure of an ostium secundum interatrial communication. The procedure led to control of strenuous exercise dyspnoea. However, in the 1 month control follow-up visit the patient reported of shortness of breath when seated and standing, with a minimum right to left shunt in the bubble echocardiogram. On physical examination, cyanosis appeared only when standing, and oxygen saturation dropped to 70% from basal 99%.

The patient was monitored for microembolic signals using transcranial Doppler sonography (TCD, videos 1 and 2) lying, sitting and standing. A platypnoea-orthodeoxia syndrome (POS) was diagnosed. The syndrome is characterised by dyspnoea that occurs in the upright position and is relieved by recumbency (platypnoea) and a significant drop in arterial oxygen saturation when moving from the supine position to the upright position (orthodeoxia). Position-dependent right-to-left shunting appears to underlie in POS.1 Contrast-enhanced transoesophageal echocardiography (cTOE, agitated saline) is the preferred diagnostic modality when intracardiac shunting is suspected. However, TCD has shown a sensitivity of 97% compared to cTOE with non-significant difference; it is the diagnostic test in the case of pulmonary arteriovenous malformations complementary to pulmonary ventilation-perfusion tests; its reported specificity is 98%.2 Effective Valsalva manoeuvre increases the sensitivity, but 10% of population may not have a temporal acoustic window. Therefore, TCD should be recommended as the first choice for screening because of its simplicity, non-invasive character, low cost and ability to assess the patient quickly in the supine as well as upright position.3

Learning points

▸ Knowledge of the patient’s history is pivotal in order to suspect platypnoea-orthodeoxia syndrome. The diagnostic test should be performed in the position that leads to dyspnoea.
▸ Transcranial Doppler with agitated saline contrast and Valsalva manoeuvre shows a sensitivity similar to contrast transoesophageal echocardiography.
▸ TCD does not require sedation and therefore allows the examination to be performed quickly in all three required positions.

Competing interests None.
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REFERENCES


Video 1 TCD monitoring (at 50 and 56 mm of depth). Right middle cerebral artery is shown on the left side of picture (red boxes) and left middle cerebral artery is on the right side (green boxes). Patient is laying (13:01:13), sitting (13:01:45) and back again to supine position at 13:01:59. "Shower embolic pattern" appears without oxygen saturation drop.

Video 2 At 13:04:29, patient was asked to stand from supine position. "Curtain pattern" (uncountable bubbles) was recorded, alongside with oxygen desaturation.