Bubbles, b ev

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Back in 1993 it had been a fairly average afternoon in the neurovascular clinic; a couple of obese, hypertensive smokers with clear-cut transient ischaemic attacks whose seemingly programmed lack of motivation to modify their vascular risk factors would subsequently challenge all the persuasive skills of my Specialist

Nurse. Well at least I could guarantee a carotid bruit for the medical students to listen to. Mixed in were the migraines without headache, or at least that is what I called them, and the paroxysmal positional vertigo that had been labelled as 'vertebrobasilar insufficiency', a term that in my view should either be used as it was first intended or discarded altogether but certainly not as an exit strategy from general practitioner surgeries or Ear, Nose and Throat clinics! On top of this a colleague's mother had been put in as an extra after an episode of amnesia and so, almost inevitably, I was running half an hour behind time with a 5 pm management meeting looming!

Then in came a lady in her early thirties who was accompanied by her husband and 10-week-old baby. The referral letter from a colleague in a nearby hospital had asked if I would discuss with her the pros and cons of closure of her patent foramen ovale (PFO). He went on to describe how 2 months earlier, in the delivery suite immediately after the birth of her daughter, she had collapsed with a dense right hemiplegia and aphasia. An immediate CT scan had shown the dense middle cerebral artery (MCA) sign along with early ischaemic changes in the left MCA territory (Fig. 1). Subsequent MR studies had reasonably excluded atheroma or carotid dissection as the cause of the stroke. The referral letter mentioned that all her basic blood tests had been

Figure 1 CT brain scan: the dense middle cerebral artery sign (arrow).

A PATIENT THAT CHANGED MY PRACTICE

Bubbles everywhere!

normal apart from a mild anaemia attributed to her pregnancy, as had a screen for vasculitic and thrombophilic disorders, an ECG and chest X-ray. There was no history of migraine. The local cardiologist had performed a transoesophageal echocardiogram and reported the prompt appearance of bubble contrast in the left atrium. After this she had been started on warfarin.

Even in 1993 this patient had brought with her a well-researched file of internet documents to discuss – not something that could be done at any speed given the residual aphasia and her daughter's crying, which defied the combined maternal experience of the clinic nursing staff. She was aware of the 20–25% background prevalence of PFO in the population, the absence of trials of antithrombotic agents, and she was adamant that she did not want open-heart surgery to close the defect. Although she knew there was no long-term outcome data after endovascular closure, it became clear that she had made up her mind that she wanted this procedure performed, not least because of her desire to have more children. I ticked off my mental check list – young stroke, no other cause identified, pro-thrombotic state in pregnancy, probable Valsalva manoeuvre at onset. Gentle probing soon revealed that she wouldn't be interested in being randomised in a trial of endovascular closure, however, she agreed to come up for a trans-cranial Doppler (TCD) ultrasound

examination because at the time I was trying out a number of different protocols for PFO detection by TCD. By this time her daughter's demand for a feed had reached a level that even I could not ignore and therefore we agreed to skip the physical examination. A quick sprint down the corridor meant the management meeting had only just started when I arrived.

A few days later the patient came to the ward for the TCD examination. On this occasion her daughter was sound asleep. Insonation of the MCA was straightforward and a good trace was obtained with relatively low gain. Using a 5 s Valsalva manoeuvre beginning at the completion of the rapid injection of agitated saline into the ante-cubital vein (Horner *et al.* 1997), the typical high intensity signals (HITS) appeared on the TCD trace and I heard the characteristic audible 'chirp' that is the most reliable method of identification of HITS (Fig. 2). The only slight oddity was that the HITS occurred between 15 and 17 s after injection rather than the more usual 10–12 s. Exactly the same timing occurred with the second injection. Slowly the penny dropped – my practice when performing TCD is to have the patient lying on a couch and I sit behind them looking down over their face. The potential significance of the telangiectasia on her lips that I must have registered subconsciously then became all too apparent and the rest, as they say, is history.

For the first time I picked the chest X-ray out of the packet – it was a typical emergency room film, taken anterior-posterior with poor penetration. The following afternoon she had a contrast-enhanced CT scan, which demonstrated several large pulmonary arteriovenous malformations (pAVM). A telephone call to her general practitioner revealed that her anaemia had predated the pregnancy. Additionally, several other members of the family who were also patients of the general practitioner had had chronic, unexplained anaemia. On subsequent detailed physical examination multiple telangiectasia were identified, firming up the diagnosis of hereditary haemorrhagic telangiectasia or Osler-Weber-Rendu disease. I then obtained the videotape of the transoesophageal echocardiogram and reviewed it with a cardiological colleague. It certainly showed bubble contrast entering the left atrium very soon after it appeared in the right atrium but, crucially, there was no unequivocal visual evidence of a PFO or for that matter an atrial septal aneurysm (ASA) as it is the combination of PFO and ASA that probably has the strongest association with recurrent 'cryptogenic' stroke (Mas *et al.* 2001).

Of course, the question about whether 'cryptogenic' ischaemic strokes in patients with pAVMs are due to paradoxical embolization is probably even more contentious than for PFOs. However, the patient's respiratory function tests demonstrated significant desaturation and therefore the intervention that was finally undertaken was coiling of the pulmonary AVMs (Fig. 3).

This salutary case convinced me to:

- Never just accept a diagnosis even from colleagues whose clinical acumen I respect. Indeed, from the outset I now tell patients who come for another opinion that it is my role to look at the evidence from a rather sceptical viewpoint, something which in general they seem to appreciate.
- Always insist on reviewing *all* the radiology personally.
- Abandon Emergency Room chest X-rays in patients with stroke as the majority are of no clinical value.
- Resist attempts to shorten consultation times.

POSTSCRIPT

Although follow up TCD could not detect any residual right-to-left shunt, I suggested that the patient should remain on low dose aspirin. Over

Figure 2 A high intensity signal (arrow) detected by transcranial Doppler in the right middle cerebral artery 17 s after the injection of agitated saline into the ante-cubital vein.

Figure 3 Pulmonary arteriovenous malformations (a) before and (b) after endovascular coiling.

the next decade she had two more children and no further strokes whilst a number of the agenda items at the management meeting in 1993 are still being discussed!

EDITOR'S NOTE

By coincidence, an article on patient foramen ovale and stroke by Jean-Louis Mas appears elsewhere in this issue.

REFERENCES

- Horner S, Ni XS, Weihs W *et al* (1997) Simultaneous bilateral contrast transcranial Doppler monitoring in patients with intrapulmonary shunts. *Journal of the Neurological Sciences*, **150**, 49–57.
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