Pulmonary hypertension: a rare but serious complication of ventriculoatrial shunts

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CASE REPORT

A 34-year-old woman presented with a 6-month history of progressive dyspnoea. Past history included cerebral palsy and epilepsy. At the age of 14 years she had a ventriculoatrial (VA) shunt for hydrocephalus secondary to congenital aqueduct stenosis. There was no past history of deep vein thrombosis, liver disease, appetite suppressant use, intravenous drug abuse or other systemic disease.

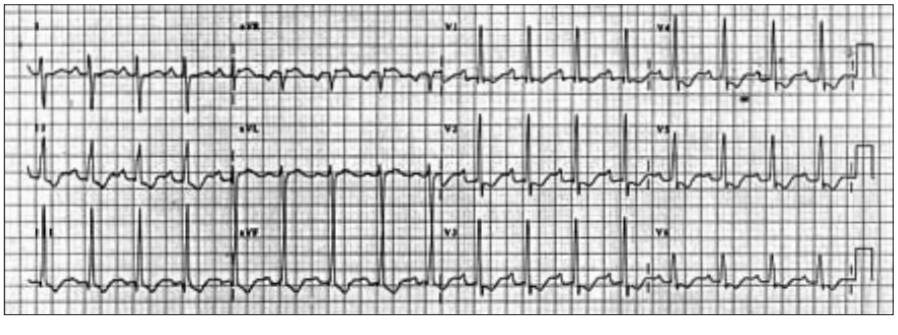
On examination she was unwell with central cyanosis, tachypnoea (respiratory rate 36 breaths per minute), sinus tachycardia (heart rate 120 beats per minute), and blood pressure 100/60 mmHg. There was ankle oedema, raised jugular venous pressure, accentuated pulmonary second sound and a right ventricular gallop.

Chest X-ray showed cardiomegaly with prominent pulmonary arteries (*Figure 1*). Electrocardiogram demonstrated right axis deviation, dominant R wave in V1 and ST depression across the precordial leads (*Figure 2*). On room air her arterial partial pressures of oxygen and carbon dioxide were 7.1 KPa and 3.2 KPa respectively. Transthoracic echocardiography showed dilated right ventricle and right atrium with severe tricuspid regurgitation and moderate pulmonary regurgitation. Estimated pulmonary artery pressure was 100 mmHg. There was no evidence of intracardiac thrombus and left ventricular systolic function normal. A ventilation perfusion scan showed multiple sub-segmental perfusion defects. Cardiac catheterization confirmed severe pulmonary hypertension (pulmonary vascular resistance 1048 dynes/sec/cm^2) with normal left heart pressures and no intracardiac shunt. Calculated cardiac index was low (1.5 litre/min/kg) with high right atrial pressure (20 mmHg) and low mixed venous oxygen saturation (50%). Pulmonary angiography and chest computed tomogram with contrast showed thrombus in the main pulmonary trunk extending into the proximal right and left pulmonary arteries. Duplex ultrasonography of both legs suggested no evidence of deep vein thrombosis. A thrombophilia, vasculitis and infection screen was negative.

The patient was commenced on nasal continuous positive airways pressure, high flow oxygen, intravenous heparin and intravenous colloid with haemodynamic stability maintained for the first 4 days. After this she became more hypoxic and hypotensive so fluids were stopped and a frusemide, dopamine and prostacyclin infusion commenced to reduce right ventricular distension with some improvement. Because of her deteriorating haemodynamic state, pulmonary thromboendarterectomy was performed with removal of extensive thrombus from the proximal pulmonary tree. The patient died in the immediate postoperative period.

Postmortem showed a distended and hypertrophied right heart with extensive chronic thromboembolic changes in the distal pulmonary tree. The tip of the VA catheter in the right atrium had no signs of infection and at this time was free of thrombus. The coroner concluded cause of death was pulmonary hypertension secondary to longstanding thromboembolic disease.

Figure 2. Electrocardiogram showing rightward axis, dominant R wave in V1, ST depression across the precordial leads.



INTRODUCTION

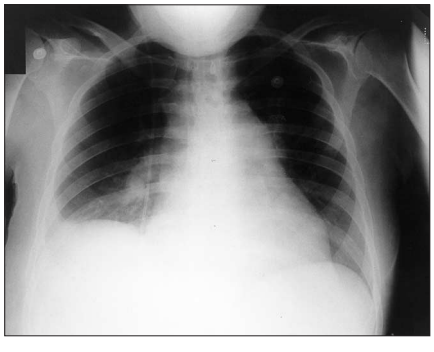
Pulmonary thromboembolic disease is a recognized and serious complication of ventriculoatrial shunts. This article describes such a case which ultimately proved fatal in a 34-year-old woman. Difficulties in the screening and management of this condition are highlighted.

DISCUSSION

Ventriculoatrial shunts for the treatment of hydrocephalus are now rarely performed because of complications such as malfunction, infection and particularly thromboembolism. Pulmonary embolism and pulmonary hypertension are recognized clinically in 0.3% and 0.4% of all cases respectively while at post mortem the frequency of these complications is much higher (59.7% and 6.3%; Drucker et al, 1984). The occurrence of pulmonary hypertension is nearly always fatal.

The onset can be early or many years later. The high incidence is not

Figure 1. Chest X-ray demonstrating cardiomegaly and prominent pulmonary arteries. The ventriculoatrial shunt can be seen in the right atrium.



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explained by a foreign body in the right atrium as patients with pacemaker leads do not have equivalent rates of thromboembolism. A possible role for the CSF being thrombogenic (in particular brain thromboplastin) and promoting pulmonary endothelial damage has been proposed (Pascual and Prakash, 1993). The risk of thromboembolism is reduced by correct positioning of the atrial catheter, ideally in the mid-atrium (Lam and Villemure, 1997). Otherwise treatment of clinically significant pulmonary hypertension is disappointing. Anticoagulation at this stage does not affect outcome. Removal of the shunt can be successful in the early course of disease but not in advanced cases.

Regular screening for pulmonary hypertension is advocated in all patients with ventriculoatrial shunts, usually with annual electrocardiogram and chest X-ray. However, both of these have only moderate sensitivity and specificity for the detection of early disease (Bossone et al, 2002). Transthoracic echocardiography is more sensitive, allows monitoring of disease progression and response to treatment (Ghio et al, 2002). If there is a suggestion of pulmonary hypertension the shunt should be replaced with a different type.

The use of prophylactic anticoagulation has not yet been proposed but could be considered in the future. Warfarin is not practical and the risks high in this patient group, many of whom have learning difficulties. Aspirin may be a safer and easier to administer alternative. **HM**

Bossone E, Paciocco G, Iarussi D, Agretto A, Iacono A, Gillespie BW, Rubenfire M (2002) The prognostic role of the ECG in primary pulmonary hypertension. *Chest* **121:** 513–18

Drucker MH, Vanek VW, Franco AA, Hansoni M, Woods L (1984) Thromboembolic complications of ventriculo-atrial shunts. *Surg Neurol* **32:** 444–8

Ghio S, Raineri C, Scelsi L et al (2002) Usefulness and limits of transthoracic echocardiography in the evaluation of patients with primary and chronic thromboembolic pulmonary hypertension. *J Am Soc Echocardiogr* **15:** 1374–80

Lam CH, Villemure JG (1997) Comparison between ventriculo-atrial and venticulo-peritoneal shunting in the adult population. *Br J Neurosurg* **11:** 43–8

Pascual JM, Prakash UB (1993) Development of pulmonary hypertension after placement of a ventriculo-atrial shunt. *Mayo Clin Proc* **68:** 1177–82

In the Public's View...

MMR: the turning point?

The *Sunday Times*' great triumph was thalidomide. Its Insight team, under editor Harold Evans, dragged the truth into the open. The newspaper cannot forget its reputation for investigative journalism, but sometimes forgets that stories have to be correct. Its 'exposures' of Debendox and of the contraceptive pill were at the least distorted if not actually incorrect. In my view, the *Sunday Times* of February 22 was back to the true tradition: its reporter Brian Deer took on the measles, mumps, rubella (MMR) story and emerged triumphant.

The *Lancet*, which published the report that started all the fuss in 1998, sprang into action. Its editor, Richard Horton, was interviewed on Radio 4's *Today* a couple of days before Deer's story was published. That interview struck me as odd. The main thrust seemed to be that Andrew Wakefield had failed to admit a conflict of interest which, if realized in 1998, would have led the *Lancet* to reject the paper. While it might be unwise or even suspicious for an author not to declare an interest, scientific papers should be accepted or rejected on scientific merit, not on interests. Horton was speaking with hindsight anyway: does he know the *Lancet* would have rejected the paper? Or would the journal have published the paper and the declaration? (And he should not have said in that interview, as he did, and repeated, that MMR was 'perfectly safe'.)

Whatever, the *Lancet* of March 6 contained nine pages of commentary and statements from just about everybody: by turns accusatory, defensive, reflective, apologetic, or affronted. Part of the commentary is a retraction – by ten authors (not Wakefield, and one could not be contacted) of the original twelve – of the interpretation that MMR caused autism. Just 6 years too late, but better than never.

The anti-MMR lobby has made much of the conflicts of interest of those in favour of the vaccine, often dismissing any and all evidence precisely and solely on those grounds. Now their champion is similarly charged, they complain of witchhunt. But there are more issues than just an alleged undeclared interest, and Wakefield has been referred to the General Medical Council for a thorough enquiry into the whole conduct of the research study, which he has welcomed. Reading through the explanation, allegation and refutation in the *Lancet*, I fear the picture after the enquiry will be just as open to interpretation as it is now. The parents who believe their children harmed will not change their minds if the General Medical Council condemns the research; it will simply be further evidence of the establishment martyring their hero. I do expect, however, even before the enquiry, that the subject will fade from public view and uptake of MMR will start increasing.

Horton has asked the government for a 'Council of Research Integrity': an independent body to investigate the conduct of research. We are in the middle of an upheaval in the way research ethics committees function, and the obvious way to police research is by an extension of their powers. But what an admission, and who would want to serve on such committees? As Horton said in an interview with the *British Medical Journal*, 'The whole system depends on trust and honour.' Sadly so, but perhaps there is too much at stake. **HM**

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