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Original Article

Cerebrovascular events in young adults after surgical repair of tetralogy of Fallot

Clara K. Chow,[1] David Amos,[1] David S. Celermajer[1,2]

[1]*Department of Cardiology, Royal Prince Alfred Hospital;* [2]*Department of Medicine, University of Sydney, Sydney, Australia*

Abstract Increasing numbers of children who have undergone intracardiac repair of tetralogy of Fallot have survived to adulthood, but often have residual haemodynamic and/or electrical abnormalities. We report the unusual observation of 4 instances of cerebrovascular accidents in young adults subsequent to surgical repair of tetralogy of Fallot.

Keywords: Stroke; congenital heart disease; cyanosis

Over the last 40 years, increasing numbers of children who have undergone intracardiac surgical repair of tetralogy of Fallot have survived to adulthood.[1,2] Many have residual cardiovascular lesions, haemodynamic and/or electrical, and require ongoing medical surveillance. Recognised late complications include pulmonary regurgitation, obstruction within the right ventricular outflow tract, dilation of the aortic root, tachy- and bradyarrhythmias, and sudden death. We currently follow-up 118 patients aged between 16 and 64 years at our hospital, a tertiary referral centre for adult congenital heart disease in New South Wales, Australia, who have undergone surgical repair of tetralogy of Fallot. Amongst these patients, we have now observed four young adults suffering cerebrovascular accidents (see Table). They are the object of our report.

Case 1

A 28-year-old man presented with weakness of the left arm and left leg, along with slurred speech, which resolved over a period of 24 hours. He was known to have tetralogy of Fallot, which had been surgically repaired when he was 6 years of age. He had been well since. He smoked 8 cigarettes per day, was overweight, with a body mass index of 32, and had elevated fasting lipids, with total cholesterol measured at 5.5 millimoles per litre, the high density fraction at 0.6 millimoles per litre, and triglycerides at 7.8 millimoles per litre. His father was alive, and suffered from diabetes mellitus. His mother had suffered a pulmonary embolism following orthopaedic surgery.

His computerised tomographic brain scan at presentation was normal. Laboratory values were unremarkable, and in particular there was no evidence of a hypercoagulable state. Holter monitoring also proved normal. Transoesophageal echo showed no evidence of intracardiac masses, thrombus, or intracardiac shunts.

His physician concluded that he had had a right brainstem infarct of uncertain aetiology. Warfarin was initially instituted and subsequently changed to aspirin.

Case 2

A 37-year-old man presented with left-sided weakness, and examination was consistent with left hemiplegic stroke. He was known to have a history of tetralogy of Fallot, which had been treated with a left Blalock-Taussig shunt at the age of 2 years, and definitive surgical repair at the age of 8 years. Review of risk factors

Correspondence to: Professor David S. Celermajer, Cardiology Department, Royal Prince Alfred Hospital, Camperdown, Sydney, NSW 2050, Australia. Tel: +61 2 951 56519; Fax: +61 2 955 06262; E-mail: david.celermajer@email.cs.nsw.gov.au

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Table. Clinical features of four patients with cerebrovascular events late after repair of tetralogy of Fallot.

	Case 1	Case 2	Case 3	Case 4
Age	28	37	39	46
Gender	Male	Male	Female	Male
Operations	Complete repair (age 6 years)	Blalock-Taussig (age 2 years) Complete repair (age 8 years)	Complete repair (age 8 years)	Blalock-Taussig (age 9 years) Complete repair (age 11 years)
Cerebral area affected	Brainstem	Right basal ganglia and thalamus	Midline pons	Right basal ganglia
Cholesterol	High	Normal	Normal	Normal
Overweight/obesity	Yes	No	No	No
Diabetes	No	No	No	Yes
Hypertension	No	No	No	No
Smoking	Yes	No	No	No
Family history of premature vascular disease	No	No	Yes	No
Other potential risk factors	Nil	Nil	Oral contraceptive pill	No
Coagulation screen	Negative	Negative	Negative	Negative
Relevant echo abnormalities	Nil	Patent foramen ovale Mildly impaired left ventricular function	Small residual ventricular septal defects	Small residual ventricular septal defects
Carotid Doppler	Not done	Not done	Normal	Normal

for cerebrovascular events proved negative, albeit that all 4 siblings had mild essential hypertension.

Computerised tomography scan showed an area of infarction involving the right side of the basal ganglia and thalamus, as well as the insular cortex and small areas of cortex of the posterior right frontal lobes. Magnetic resonance imaging performed at a later date showed evidence of previous infarction in the distribution of the lentriculostriate branches of the middle cerebral artery. Transthoracic echo showed mildly impaired left ventricular contractility, a moderately dilated and mildly impaired right ventricle, severe pulmonary regurgitation, but no evidence of intracardiac thrombus. A bubble study was initially inconclusive, but subsequent studies demonstrated a patent oval foramen, with a small right-to-left shunt during the Valsalva manoeuvre.

Laboratory examination revealed no abnormalities; in particular there was no evidence of a hypercoagulable state. It was concluded that he had suffered an infarct in the territory of the middle cerebral artery secondary to a cardioembolic event. He was commenced on Warfarin. Subsequent to this presentation, operation for repair of his severe pulmonary regurgitation confirmed patency of the oval foramen, which was repaired. His Warfarin was stopped prior to operation, and was not recommenced.

Case 3

A 39-year-old woman presented with acute diplopia, with associated loss of vertical down gaze and hypophonia. Concurrently she felt very tired, and went to sleep for about 2 hours. When she woke, her symptoms had resolved. At review, neurological examination was normal.

She was known to have a past history of tetralogy of Fallot, having undergone surgical repair at the age of 8 years. She had been well since then. A review for risk factors for cerebrovascular disease, including obesity, use of tobacco, hyperlipidaemia and hypertension, was negative. Her father had had a history of ischaemic heart disease, and had undergone coronary arterial grafting. Her only medication was the oral contraceptive pill. Computerised tomography and magnetic resonance imaging demonstrated a cerebral infarct in the medial portion of the left thalamus. Routine tests of blood revealed no abnormalities, and a hypercoagulability screen was negative. Carotid and venous Doppler scans also carried out to determine the aetiology of the stroke proved negative. Transoesophageal echo showed a small persistent ventricular septal defect, with left to right flow, normal left ventricular size and function, a mildly dilated and hypertrophied right ventricle with mild obstruction of the right ventricular outflow tract, and no intracardiac thrombus.

It was concluded that the history of the episode suggested an event occurring in the midbrain region of the brainstem, and was consistent with a transient ischaemic attack. The old lacunar infarct found on magnetic resonance imaging was deemed to be a separate event. Her oral contraception was stopped, and she was commenced on aspirin.

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Case 4

A 46-year-old man presented with sudden onset of left sided weakness, falling to the left, dysarthria, and visual disturbances. Magnetic resonance imaging one day later revealed acute infarction of the right basal ganglia, together with accumulation of a number of focal areas of blood in the right parietal lobe, of uncertain significance. Due to concern that these could be related to bleeding associated with tiny vascular malformations, we also performed computerised tomography of the brain and computerized tomographic angiography of the Circle of Willis, which failed to reveal any areas of haemorrhage in the right parietal lobe, nor any additional vascular abnormalities.

He was known to have a history of repaired tetralogy of Fallot and multiple ventricular septal defects. A right Blalock-Taussig shunt had been constructed at the age of 9 years, with complete repair undertaken at the age of 11years. The multiple ventricular septal defects could not be all closed at time of complete repair, but were thought to be of no haemodynamic significance.

In terms of cerebrovascular risk factors, type II diabetes had been diagnosed 3 years previously, and was well controlled on a single oral hypoglycaemic agent. Aside from this, he was a non-smoker, with no family history of vascular disease, no hypertension, nor hyperlipidaemia. Past history included a history of forearm melanoma excised some 10 years ago, and benign prostatic disease. Further investigation for stroke included Doppler interrogation of the carotid arteries, which was normal, a transthoracic echocardiogram, which showed mild to moderate dilation of the left ventricle with good systolic function, intermediate motion of the septal wall, reasonable right ventricular size and function, mild residual obstruction of the right ventricular outflow tract, no pulmonary regurgitation, and small residual ventricular septal defects. A procoagulant screen was unremarkable.

Discussion

At our hospital, a tertiary referral centre for adults with congenital heart disease in New South Wales, Australia, we currently follow-up 118 patients aged between 16 and 64 years subsequent to surgical repair of tetralogy of Fallot, suggesting a prevalence of cerebrovascular accidents in this highly selected group of up to 1 in 30. This is higher than the rate of stroke in the Australian population. The North East Melbourne Stroke Incidence Study, known as the NEMESIS study, found the attack rate for stroke in males and females between the ages of 25 and 44 years old was 36 in 100,000.[3] This suggests that a history of previous tetralogy of Fallot repair may be a risk factor for early stroke. If so, these patients may need to have their vascular risk factors more closely monitored, as well as being considered for prophylactic treatment with aspirin as adults.

Investigation of stroke in young people usually involves assessment of vascular risk factors including smoking, diabetes, cholesterol, obesity, hypertension and family history, as well as additional tests of blood coagulation coupled with echocardiography.

A case report published in 2002 describes a patient suffering a stroke of the vertebrobasilar territory as a late complication of a Blalock-Taussig shunt due to a subclavian steal phenomenon.[4] Otherwise, as far as we are aware, cerebrovascular accidents have not been reported previously in this population, nor as a prevalent complication in other forms of repaired cyanotic heart disease, although stroke is a recognised complication of unrepaired cyanotic heart disease.[5,6]

We have encountered, therefore, a small series of patients with surgically repaired tetralogy of Fallot who have suffered cerebrovascular events at a young age. Some of them had co-existing risk factors that might explain these observations. Indeed, the mechanism may relate to residual intracardiac communications demonstrated in three of the four cases. Further surveillance of survivors with repaired cyanotic congenital heart disease may be informative regarding the true prevalence of this rare yet dangerous late complication.

References

1. Murphy JG, Gersh BJ, Mair DD, et al. Long-term outcome in patients undergoing surgical repair of tetralogy of Fallot. N Engl J Med 1993; 329: 593–599.
2. Katz NM, Blackstone EH, Kirklin JW, Pacifico AD, Bargeron LM. Late survival and symptoms after repair of tetralogy of Fallot. Circulation 1982; 65: 403–410.
3. Thrift AG, Dewey HM, Macdonell RA, McNeill JJ, Donnan GA. Stroke incidence on the east coast of Australia. The North East Melbourne Stroke Incidence Study (NEMESIS). Stroke 2000; 31: 2087–2092.
4. Gailoud P, Hillis A, Perler B, Murphy KJ. Vertebrobasilar stroke as a late complication of a Blalock-Taussig Shunt. Ann Neurol 2002; 52: 231–234.
5. Berthrong M, Sabsiton DC. Cerebral lesions in congenital heart disease: a review of autopsies on one hundred and sixty-two cases. Bull Hopkins Hospital 1951; 89: 384–401.
6. Phornphutkul C, Rosenthal A, Nadas AS, Berenberg W. Cerebrovascular accidents in infants and children with cyanotic congenital heart disease. Am J Cardiol 1973; 32: 329–334.