

# Fatal pneumococcal septicaemia associated with asplenia and isomerism of the right atrial appendages

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## Abstract

**A 22 month old male infant with isomerism of the right atrial appendages (associated with a complete atrioventricular septal defect, double outlet right ventricle, and pulmonary valve stenosis) died from pneumococcal septicaemia after a 36 hour illness. He had not been given penicillin prophylaxis. Asplenia was confirmed at necropsy.**

**In a survey of 50 paediatric cardiologists in the United Kingdom, 13 (33%) of the 40 (80%) who replied did not advise any prophylactic measures against pneumococcal infection in patients with isomerism of the right atrial appendages.**

Isomerism of the right atrial appendages is usually found in association with severe forms of cyanotic congenital heart disease.<sup>1</sup> It is almost always associated with asplenia.<sup>2</sup> This may predispose the child to infection by *Streptococcus pneumoniae*, *Neisseria meningitidis*, and *Haemophilus influenzae*.<sup>3</sup>

Daily prophylactic penicillin and vaccination with a pneumococcal vaccine (Pneumovax) have both been shown to reduce the incidence of pneumococcal infection in patients with asplenia.<sup>4,5</sup>

We report this case to highlight the continued need for awareness of the increased infection risk in children with isomerism of the right atrial appendages. This case also prompted us to send a questionnaire to 50 paediatric cardiologists in the United Kingdom to see what were their current recommendations for prophylaxis in children with this type of defect. Forty (80%) replied and the results are reported.

## Case report

A male infant was noted to be cyanosed at birth. Having failed a hyperoxia test he was transferred for cardiac assessment when he was one day old.

Physical examination showed a cyanosed infant weighing 4.0 kg with a 3/6 ejection systolic murmur that was loudest at the second left intercostal space. An echocardiogram showed isomerism of the right atrial appendages. The pulmonary veins drained to the left sided atrium. There was a complete atrioventricular septal defect with a double outlet right ventricle. In addition, there was pulmonary valve stenosis with a Doppler estimated gradient of 36 mm Hg.

His cyanosis worsened and when he was 10 days old a modified right Blalock-Taussig shunt was constructed with a 5 mm polytetrafluoroethylene (Gore-tex) graft. The post-operative course was uneventful. He was discharged. He remained well and continued to thrive. No prophylactic penicillin was advised.

When he was 22 months old, after a short 6 hour febrile illness he became lethargic and stuporose. He was admitted to the local hospital where a full screen, including a lumbar puncture and a urine screen for pneumococcal antigens, was performed.

Treatment with intravenous benzylpenicillin (50 mg/kg four hourly) and gentamicin (3 mg/kg eight hourly) was started. Six hours later there was further deterioration in his level of consciousness and he was transferred to the regional paediatric intensive care unit.

On arrival he was hypotensive with a systolic blood pressure of 50 mm Hg and with a temperature gap of 10°C between the core and periphery. He was given colloid and treatment with dobutamine (10 µg/kg/min) and dopamine (3 µg/kg/min) was started. Initial haematological investigations showed a normal white cell count ( $6.7 \times 10^9/l$ ) and evidence of disseminated intravascular coagulation.

Echocardiography showed that the Blalock-Taussig shunt was patent and no intracardiac vegetations were seen. Ventricular function was also normal. Blood cultures taken at the local hospital isolated *Streptococcus pneumoniae* at 24 hours; the organism was fully sensitive to penicillin.

Despite treatment, bradycardia and hypotension developed after 36 hours of illness and he died. At necropsy no vegetations were seen in either the heart or on the shunt and no splenic tissue was found.

## Results of survey

Forty (80%) paediatric cardiologists responded to the survey. Half of the respondents recommended daily penicillin and pneumococcal vaccination in children with isomerism of the right atrial appendages. Four (10%) recommended penicillin alone and three (7%) recommended only pneumococcal vaccination. Thirteen (33%) recommended neither.

Twenty one of the 24 (60%) who recommended penicillin prophylaxis would have started it at the time of diagnosis. Three consultants would have waited until the child was 6-12 months old before starting prophylaxis. Twenty two recommended its indefinite use,

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one suggested stopping it at eight years of age, and the other immediately after pneumococcal vaccination, which was at three years of age in their unit.

Twenty three (57%) recommended pneumococcal vaccination as part of their prophylaxis policy, and 20 would have combined it with penicillin. One would have stopped penicillin on vaccination and two recommended pneumococcal vaccination alone. The timing of the pneumococcal vaccination was variable. About half (12 paediatric cardiologists) suggested vaccination at two years of age, two (10%) at three years, two (10%) at some time between the age of six to twelve months, and the remaining seven (30%) left it to the discretion of the local paediatrician. One consultant also recommended that the *Haemophilus influenzae* and meningococcal vaccines should also be given.

### Discussion

About 1% of infants with congenital heart disease have isomerism of the right atrial appendages.<sup>1</sup> In life this is usually diagnosed when echocardiography shows that the aorta and inferior vena cava are positioned on the same side of the spine.<sup>6</sup>

Isomerism of the right atrial appendages is almost always associated with asplenia.<sup>2</sup> A review of 52 patients with asplenia and congenital heart disease by Waldman *et al* showed that most died of their associated congenital heart defect in the first year, usually in the first month; after this period the commonest cause of death was infection.<sup>7</sup> As surgical techniques improve, more children with isomerism of the right atrial appendages may survive longer and the risk of overwhelming infection will be high.<sup>3</sup>

Recent publications of guidelines for the use of pneumococcal vaccination<sup>8,9</sup> have highlighted the importance of prophylaxis against

pneumococcal infection in patients with functional asplenia and those undergoing splenectomy, in whom prophylactic penicillin is already known to reduce the incidence of pneumococcal infection.<sup>4</sup> But no attention has been paid to patients with asplenia associated with isomerism of the right atrial appendages. This is clearly reflected by a variable policy of recommendations by the paediatric cardiologists in the United Kingdom.

We believe that these guidelines are applicable to children with isomerism of the right atrial appendages. At diagnosis daily penicillin prophylaxis should be started and continued indefinitely. The child should also be vaccinated at the age of two with Pneumovax II, and this should be repeated indefinitely every 6 years. Finally, the importance of the susceptibility of these patients to life threatening infection should be emphasised to the child's parents, general practitioner, and general paediatrician.

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