

## Q fever tricuspid valve endocarditis

J M Lupoglazoff, P Brouqui, S Magnier, U Hvass, A Casasoprana

### Abstract

**Q fever is a zoonosis caused by *Coxiella burnetii*. The most frequent clinical expression of the chronic form is a bacterial culture negative aortic or mitral endocarditis. A case of tricuspid valve endocarditis due to *C burnetii* is described, with a favourable outcome after treatment with doxycycline and hydroxychloroquine.**

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Q fever is a disease caused by the rickettsia *Coxiella burnetii*.<sup>1</sup> This strict intracellular pathogen parasitises the mononuclear cells. We present what we believe to be the first reported case of Q fever tricuspid valve endocarditis.<sup>1,2</sup>

### Case report

An 8 year old girl with a history of persistent high grade fever was referred with the diagnosis of tricuspid endocarditis. She presented with hepatosplenomegaly, a continuous cardiac murmur on the right side of the sternum, and signs of right cardiac failure. An echocardiogram showed dilated right cardiac cavities and a huge vegetation of the tricuspid valve. The right coronary artery was dilated with a fistula between the right coronary artery and the right ventricle; the left coronary artery was normal.

None of the six blood cultures taken before antibiotic treatment was positive. Serum *C burnetii* antibodies were positive by microimmunofluorescence assay at a titre of 800, 800, and 0 for phase I IgG, IgM, and IgA respectively, and 3200, 1600, and 0 for phase II IgG, IgM, and IgA respectively.<sup>3</sup> Treatment with 200 mg/day of doxycycline and 600 mg/day of hydroxychloroquine resulted in defervescence by 48 hours. Medical treatment did not control cardiac failure and so valve replacement became necessary. The aortogram taken before surgery showed a voluminous fistula between the right coronary artery and the ventricular face of the tricuspid valve. The fistula was repaired and the valve replaced by a cryopreserved mitral valve homograft. Postoperative follow up was uncomplicated. Microbiological analysis of the vegetation was negative for bacteria and fungi, but citrate synthase gene amplification and subsequent DNA sequence analysis showed the presence of *C burnetii* DNA in the ablated vegetation. The child recovered rapidly and was discharged taking doxycycline and hydroxychloroquine for a planned two years.

### Discussion

Q fever presents variably.<sup>1</sup> The acute form is characterised by pneumonitis, granulomatous hepatitis, or isolated fever. Bacterial culture negative endocarditis is seen in the chronic form, and mainly involves damaged aortic or mitral valves.<sup>2</sup> We could find no previous report of tricuspid valve endocarditis due to *C burnetii*. One possible explanation for our case is that the spray which gushed from the congenital coronarocardiic fistula might have injured the septal valve endothelium allowing *C burnetii* to engraft and cause endocarditis. The valves are usually severely damaged by *C burnetii*,<sup>2</sup> and distal embolism in the central nervous system is a frequent initial clinical manifestation of Q fever endocarditis.<sup>2</sup> Culture of removed valves can help isolate the infectious pathogen despite antibiotic treatment.<sup>4,5</sup> Unfortunately, neither blood culture nor culture of the removed valve was attempted in our case. Nevertheless, *C burnetii* serology is very specific. In patients with endocarditis, a titre of 1:800, or greater, of IgG directed against phase I antigen has a positive predictive value of 98% and a sensitivity of 100%.<sup>3</sup> Identification of *C burnetii* DNA in tissue by polymerase chain reaction also strongly supports a diagnosis of Q fever.<sup>3</sup> *C burnetii* specific citrate synthase DNA sequence detection in the vegetation, when added to the serological data, confirmed the diagnosis.

Treatment of Q fever endocarditis is based on a prolonged regimen of antibiotics. *C burnetii* is an obligate intraleucocytic Gram negative organism, which explains the lack of efficacy of most antibiotics. The low pH of phagolysosome inhibits the bactericidal effect of doxycycline, which is necessary to eradicate the organism, particularly in immunosuppressed patients.<sup>2</sup> The addition of a lysosomotropic agent such as hydroxychloroquine, which raises the pH of vacuoles, restores the bactericidal activity of doxycycline.<sup>6</sup> Preliminary results of treating Q fever endocarditis with the combination of doxycycline and hydroxychloroquine have been successful.<sup>6</sup> While the recommended duration of treatment with doxycycline and quinolones is at least three years,<sup>2</sup> with numerous relapses, a two year treatment with no relapse at prolonged follow up seems possible. Although cyclines are contraindicated in children, it remains the only effective available treatment.

Q fever endocarditis is not a rare disease and is found world wide.<sup>1,2</sup> In case of fever with negative blood culture, Q fever should be suspected and the prescription of the specific serology performed. Prolonged treatment with

Service de Cardiologie  
Pédiatrique,  
Hôpital Robert Debré,  
Paris, France  
J M Lupoglazoff  
S Magnier  
A Casasoprana

Unité des Rickettsies,  
CNRS UPRES,  
Marseille, France  
P Brouqui

Service de Chirurgie  
Cardiovasculaire,  
Hôpital Bichat,  
Paris, France  
U Hvass

Correspondence to:  
Dr J M Lupoglazoff,  
Service de Cardiologie,  
Hôpital Robert Debré,  
48 Boulevard Serurier,  
75019 Paris, France.

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doxycycline combined with hydroxychloroquine is recommended.

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