

# Unusual evolution of a non-hacek Gram-negative endocarditis in a patient with Turner syndrome

CARINE BECH<sup>1</sup>, BENEDIKT HUTTNER<sup>2</sup>, JUDITH ARCHAMBAULT PATTERSON<sup>1</sup>, JEAN-JACQUES PERRENOUD<sup>1</sup>

<sup>1</sup>Department of Rehabilitation and Geriatrics, University Hospital of Geneva, CH-1226 Thônex, Geneva, Switzerland

<sup>2</sup>Infection Control Program, University of Geneva Hospitals and Medical School, CH-1211 Geneva 14, Switzerland

Address correspondence to: Carine Bech. Tel: +41 22 718 45 90; Fax: +41 22 718 45 85. Email: Carine.Bech@hcuge.ch

## Abstract

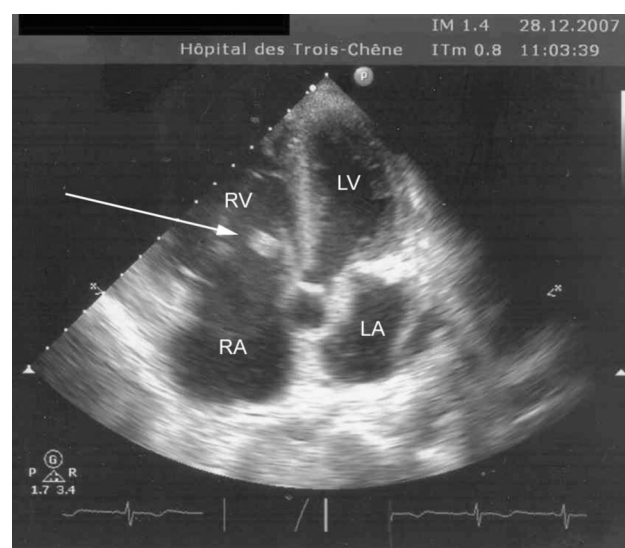
Non-HACEK Gram-negative endocarditis is a rare but severe illness, and the diagnosis can be difficult to establish. Here, we report the case of a 72-year-old woman with Turner syndrome suffering from non-typhoid *Salmonella* endocarditis of the tricuspid valve, who benefited from prompt antibiotic treatment allowing a quick and complete recovery.

**Keywords:** non-HACEK Gram-negative endocarditis, Turner syndrome, elderly

## Case report

A 72-year-old non-institutionalised woman was hospitalised for evaluation by her primary care physician for cough, expectoration and dyspnoea of 1 week's duration before admission. Her medical history was significant for Turner syndrome associated with mental retardation and congenital cardiac abnormalities (thickening of the tricuspid valve and discrete tricuspid insufficiency). The patient denied fever, chills, diarrhoea or vomiting and had not recently travelled. On admission, her temperature was 38.0°C and physical examination revealed a mesocardial systolic heart murmur (2/6) and peripheral oedema. Lung and abdominal findings were unremarkable. Blood analysis showed leucocytosis (11.2 G/l) with 2% band forms, elevated C-reactive protein (221 mg/l) and elevated liver function tests (ASAT 119 mmol/l, ALAT 46 mmol/l, GGT 173 mmol/l, bilirubin 12 µmol/l). Chest radiography was normal. An ECG showed a right bundle branch block.

Two sets of blood cultures taken on admission yielded *Salmonella enterica serovar enteritidis*. A trans-thoracic echocardiogram done for investigating right cardiac failure confirmed the thickening of the tricuspid valve and revealed a vegetation on its posterior leaflet (Figure 1), with a moderate-to-severe tricuspid insufficiency as well as significant dilatation of the right cavities with moderate arterial pulmonary hypertension (50 mmHg). A computerised tomography (CT) scan of the abdomen revealed no aneurysms, and an HIV test was negative. A diagnosis of infective endocarditis was made based on one major and three minor modified Duke's criteria [1]: evidence of endocardial involvement on the echocardiogram, predisposing heart condition, fever and microbiological evidence. The patient was immediately



**Figure 1.** Two-dimensional echocardiography: apical four-chambers view. Voluminous vegetation on the posterior tricuspid leaflet (arrow).

treated with ceftriaxone (1 g IV per day), and the evolution was rapidly favourable. Two weeks later, a trans-thoracic echocardiogram showed a drastic diminution of the size of the vegetation. The patient was discharged after 6 weeks of intravenous antibiotic therapy. Six months later, she is well and without complications.

## Discussion

To our knowledge, this is the first documented case of non-HACEK Gram-negative endocarditis in a 72-year-old

patient with Turner syndrome. Cardiovascular complications, notably aortic dissection, are an important cause of morbidity and mortality in patients with Turner syndrome (monosomy for the X chromosome) [2]. Endocarditis, however, seems to be an infrequent complication, despite a high prevalence of bicuspid aortic valves in affected patients [3]. Gram-negative bacteria other than the HACEK organisms are a rare cause of endocarditis in general. *Salmonella* species display a well-known propensity for the vascular endothelium, and bacteraemia with these organisms is often complicated by mycotic aneurysms, notably in the elderly [4]. *Salmonella* species also represent a large proportion of cases of non-HACEK Gram-negative endocarditis described in the literature [5]. For decades, non-HACEK Gram-negative bacillus endocarditis has been reported mainly among drug users. A recent international collaborative study analysing 2,761 cases of endocarditis identified 49 cases of endocarditis due to non-HACEK Gram-negative bacilli (1.8% of all cases). Only one of them was caused by *Salmonella* species. Most of these non-HACEK endocarditis were associated with health-care contact (59% of the patients had a prosthetic valve) [6].

It is noteworthy that, similar to our patient's case, most cases of bacteraemia caused by *Salmonella* species are not accompanied by gastrointestinal symptoms [7]. Except for her advanced age, our patient did not present with any of the known risk factors for *Salmonella* bacteraemia such as diabetes, treatment with immunosuppressive drugs, HIV or neoplasm. In addition, the right-sided localisation of the vegetation and the absence of a prosthetic valve might explain the favourable outcome in our patient, which contrasts with the bad prognosis associated with this kind of disease described in the literature [5]. The optimal choice and duration of antibiotic therapy for non-HACEK Gram-negative endocarditis are unclear, but cardiac surgery in combination with prolonged courses of antibiotic therapy is often necessary [8].

## Conclusion

Endocarditis caused by bacteria not commonly associated with this pathology, like non-HACEK Gram-negative bacilli, can be difficult to diagnose and outcome can be poor. A high index of suspicion is necessary, and it should be kept in mind that this disease is not limited to classical risk groups like injection drug users. This case also illustrates that, in the absence of immuno-deficiency or prosthetic material, prompt and adequate antibiotic treatment can lead to a favourable

outcome even in an older patient with underlying cardiac abnormalities.

## Key points

- Non-HACEK Gram-negative endocarditis is a rare but severe illness.
- In the absence of immuno-deficiency or prosthetic material, prompt and adequate antibiotic treatment can lead to a favourable outcome.

## Acknowledgement

We thank Prof. J. P. Michel for the revision of the manuscript.

## Conflicts of interest

There are no conflicts of interest.

## References

1. Li JS, Sexton DJ, Mick N *et al.* Proposed modifications to the Duke criteria for the diagnosis of infective endocarditis. *Clin Infect Dis* 2000; 30: 633–8.
2. Bondy CA. Congenital cardiovascular disease in Turner syndrome. *Congenit Heart Dis* 2008; 3: 2–15.
3. Sachdev V, Matura LA, Sidenko S *et al.* Aortic valve disease in Turner syndrome. *J Am Coll Cardiol* 2008; 51: 1904–9.
4. Benenson S, Raveh D, Schlesinger Y *et al.* The risk of vascular infection in adult patients with nontyphi *Salmonella* bacteremia. *Am J Med* 2001; 110: 60–3.
5. Aubron C, Charpentier J, Trouillet JL *et al.* Native-valve infective endocarditis caused by enterobacteriaceae: report on 9 cases and literature review. *Scand J Infect Dis* 2006; 38: 873–81.
6. Morpeth S, Murdoch D, Cabell CH *et al.* Non-HACEK Gram-negative bacillus endocarditis. *Ann Intern Med* 2007; 147: 829–35.
7. Gordon MA. *Salmonella* infections in immunocompromised adults. *J Infect* 2008; 56: 413–22.
8. Baddour LM, Wilson WR, Bayer AS *et al.* Infective endocarditis: diagnosis, antimicrobial therapy, and management of complications: a statement for healthcare professionals from the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease, Council on Cardiovascular Disease in the Young, and the Councils on Clinical Cardiology, Stroke, and Cardiovascular Surgery and Anesthesia, American Heart Association: endorsed by the Infectious Diseases Society of America. *Circulation* 2005; 111: e394–e434.

Received 7 November 2008; accepted in revised form 11 February 2009