

Ebstein's disease discovered after the age of 50 years

Clinical Case Portal

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Case Report

This case illustrates an unusually late discovery of Ebstein's disease with a concomitant appearance of a severe conduction defect.

The main therapeutic option in the present case was dual chamber pacing.

Patient: 62-year old man. Congenital pulmonary stenosis diagnosed at age 20. At the age of 52, after some 10 years of treatment for asymptomatic systemic hypertension, he had a second cardiological examination, and Ebstein's disease was diagnosed.

Patient history prior to current observation :

A 62 years old technician had undergone cardiac catheterisation at age 20, apparently because of unusual auscultation findings. Congenital pulmonary stenosis was diagnosed. At the age of 52, after some 10 years of treatment for asymptomatic systemic hypertension, he experienced shortness of breath and rapid

and apparently regular palpitations that motivated a second cardiological examination. At this time, his general condition was good.

Clinical findings on admission, evolution and outcome :

Auscultation revealed a widely split first heart sound and a soft systolic murmur. He was in sinus rhythm fig. 1.

M-mode echocardiography fig. 2 showed the classical delay of tricuspid closure, a large tricuspid valve and an enlarged right ventricle, permitting the definite diagnosis of Ebstein's anomaly. No further treatment was proposed.

Nearly 9 event-free years later, at the age of 61, during a period of heavy professional overload, he experienced recurrence of dyspnoea and palpitations and also several syncopal attacks (5 to 6 episodes). On admission to hospital he was groggy but well orientated. His neurological status was normal, the oxygen saturation was 82% and the partial oxygen pressure under FIO₂ of 50% was 68 mmHg. Clinical examination was unchanged and he was still in sinus rhythm. Holter monitoring and exercise tests were also normal. A 7 day rhythm monitoring fig. 3 (R-test) showed an asymptomatic, short, non-sustained ventricular tachycardia of 4 complexes.

Transthoracic echocardiography fig. 4, fig. 5, fig. 6, fig. 7, fig. 8, fig. 9) showed the classical features of Ebstein's anomaly with a huge abnormal tricuspid valve, right ventricular enlargement and a large area of atrialised ventricle. An inter-atrial septal aneurysm was present. Contrast echocardiography confirmed the suspected right-to-left shunt.

The diagnosis of severe Ebstein's anomaly with an aneurysm of the inter-atrial septum and a bidirectional shunt was established. However, the episodes of syncope and pre-syncope remained to be explained. Therefore, a search was made for an arrhythmia and/or conduction defect.

Electrophysiological studies fig. 10 revealed the clue to the syncopal episodes: an infra-Hissian second degree AV block which is invisible on the simultaneous surface ECG.

A dual-chamber pacing system was inserted and the patient remains physically well 1 year after pacemaker insertion. He continues to decline any surgical intervention such as a Glenn-shunt (superior vena cava to right pulmonary artery) + closure of the atrial septal defect.

A rather disturbing result: an infra-Hissian second degree AV-block is easily visible, especially on the 4th tracing where a Hissian signal follows every atrial signal but a ventricular signal is present only after every second Hissian signal. Notice, that this 2nd degree AV-block is absolutely not visible on the simultaneous surface ECG

(Notice also the different recording speeds. RR — interval is of 950 ms on the surface ECG, AA-interval is of 475 ms on the intracardiac recordings. This tracing was recorded under Isuprenalin-infusion).

Discussion

This case illustrates an unusually late discovery of Ebstein's disease with a concomitant appearance of a severe conduction defect, which is a rather rare observation (1), ventricular pre-excitation syndromes and atrial arrhythmias being more frequent in these cases (2).

Ebstein's disease has a large clinical spectrum. In asymptomatic and mildly symptomatic patients, medical management is recommended. Progressive clinical deterioration with functional class NYHA III or IV, cardiomegaly, cyanosis and/or arrhythmias are important reasons to consider surgical intervention. (3).

Tricuspid valvuloplasty is currently the option favoured by many authors (4,5).

Complementary and/or alternative options are tricuspid valve replacement by a bio-prosthesis, partial bi-

ventricular repair with reduction of right ventricular volume overload (6) and closure of the very frequently associated atrial septal defect.

The main therapeutic option in the present case was obviously dual chamber pacing. Permanent pacing in patients with Ebstein's disease is a challenge due to anatomical variations (and tricuspid valve replacement). In a large Mayo Clinic series of 401 patients with Ebstein's disease, 15 (3.7%) required permanent pacing. Amongst them, 11 had AV block and 4 had sinus node dysfunction (1).

Only two of them underwent a "normal" transvenous dual chamber pacing. Epicardial leads and mixed systems were very often required. 27% of them had complications requiring surgical intervention (1).

Conclusion

Therefore, the present patient seems to be an exception regarding at least two different aspects:

His congenital malformation has been recognised unusually late in his life. Evens so, his condition did not absolutely necessitate surgical treatment (which he had declined any way...).

The unusually late appearance of a severe AV conduction defect, which is rather rare in Ebstein's disease, and which could be treated by "normal" trans-venous dual chamber pacing without short term complications.

References

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Fig. 1 :
[Ebstein's disease 12-lead surface ECG](#)



Fig. 2 :

[2D-derived M-mode echocardiography](#)

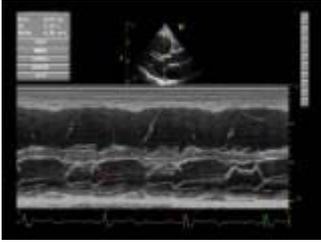


Fig. 3 :

[Ebstein's disease R-test \(= 7 days rhythm monitoring\)](#)



Video 1 :

[Ebstein's disease TTE: 4-chamber view-I](#)



Video 2 :

[Ebstein's disease 4-chamber view-II](#)



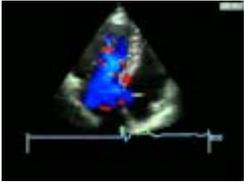
Video 3 :

[Ebstein's disease TTE: short axis view](#)



Video 4 :

[Ebstein's disease TTE: 4-chamber view, colour Doppler](#)



Video 5 :

[TTE: 4-chamber view, contrast-I](#)



Video 6 :

[TTE, 4-chamber view, contrast II](#)



Fig. 4 :

[Ebstein's disease Electrophysiological study. Simultaneous 12-lead surface ECG](#)

