RECURRENT ECTOPIC JUNCTIONAL TACHYCARDIA IN EBSTEIN'S ANOMALY

— Case Report of a 67-year-old Man —

KOSHI MATSUYAMA, M.D., YUTAKA HORIO, M.D., HIDEO UCHIDA, M.D.*
MANABU ROKUTANDA, M.D., KEN OKUMURA, M.D.*, KYOJI TAKAOKA, M.D.
NOBUYA IMOTO, M.D., YOSHIHIRO KIMURA, M.D.*, SHUKURO ARAKI, M.D.*
and HIROFUMI YASUE, M.D.

We reported the case of a 67-year-old man with Ebstein’s anomaly, the oldest patient with this disease in Japan as far as we know. His condition was often accompanied by ectopic junctional tachycardia with isorhythmic AV dissociation, which made him complain of palpitation by elevating right intra-atrial pressure. Although enhanced automaticity was the most likely mechanism of this tachycardia, it was abruptly initiated and terminated by a single premature contraction. Because this interesting character can not be explained solely by the usual ideas of automaticity, we suggest that ectopic junctional tachycardia in the present case may somewhat resemble the phenomenon of triggered automaticity.

ECTOPIC junctional tachycardia is characterized by enhanced automaticity in the atrio-ventricular (AV) junction with a modest increase in ventricular rate (70–130/min) and by a lack of abrupt onset and/or termination.1,2 Thus, this arrhythmia is usually termed non-paroxysmal atrio-ventricular junctional tachycardia (NPJT)1,2 or accelerated junctional tachycardia, and it typically occurs in the clinical settings of digitalis intoxication, acute diaphragmatic infarction, acute rheumatic fever, and following open heart surgery.1–3 Ectopic junctional tachycardia in Ebstein’s anomaly is considered extremely rare3 and has not been reported in detail as far as we know.

In this study, we describe a 67-year-old man who is believed to be the oldest patient with Ebstein’s anomaly yet reported in Japan, and whose condition was accompanied by recurrent ectopic junctional tachycardia which showed abrupt onset and termination.

CASE REPORT

A man, aged 67, was admitted to Kumamoto University Hospital in February 1983 complaining of palpitation and a feeling of “pressure” with abrupt onset and termination in the anterior chest. He was asymptomatic until 1956 when at age 40 he experienced episodes of palpitation briefly about once a month. Thereafter, the frequency and duration gradually increased until he felt precordial oppression for several hours several times a day by the time of admission. The patient had not contracted any other signifi-

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Division of Cardiology and *First Department of Internal Medicine, Kumamoto University School of Medicine, Kumamoto, Japan
Mailing address: Koshi Matsuyama, M.D., Division of Cardiology, Kumamoto University School of Medicine, 1-1-1 Honjo, Kumamoto 860, Japan

Japanese Circulation Journal Vol. 49, April 1985
AV Junctional Tachycardia in Ebstein’s Anomaly

Fig.1. Chest X-ray film.

cant disease, nor had he been taking any con-
tinuous medication. There was nothing to note
in his personal or family history.

Physical examination revealed a moderately
developed man without acute distress. Height
was 160.4 cm, body weight 53.5 kg, and body
temperature 36.5°C. Blood pressure was 100/62
mmHg, and pulse was 64/min with regular
sinus rhythm. Mild flush was found on both
cheeks. Cyanosis, clubbing, and edema were
absent. The cervical veins were not distended,
and there was no goiter. The first and second
heart sounds split widely, and the fourth sound
was found. A grade 3/6 holosystolic murmur,
so-called “scratchy” murmur, was heard most
strongly along the left lower sternal border.
Lungs were clear on percussion and auscultation.
Hepatosplenomegaly was not observed. There
were no additional findings to note in physical
examination.

There was a mild polycythemia; red blood cell
count was 560 x 10^6/mm³, hemoglobin 17.2
g/dl, and hematocrit 51.6%. Other hemogram
tests, urinalysis, and blood chemical surveys
were all within normal range.

Chest X-ray disclosed mild cardiomegaly
(CTR = 56%) and a somewhat globular appearance
to the overall cardiac silhouette. Relative pulmo-
mary hypovascularity was also demonstrated
(Fig. 1).

The electrocardiogram recorded when the
patient felt no palpitation showed a regular sinus

![Control and Palpitation Attack ECGs](image)

Fig.2. Standard electrocardiographic recordings at the control state (left panel) and at
the palpitation attack (right panel).
The lowermost illustrations are the continuous recordings in lead I.

*Japanese Circulation Journal* Vol. 49, April 1985
Fig. 3. Intracavitary electrocardiograms at the sinus rhythm (upper panel) and at the ectopic junctional tachycardia (lower panel).

Shown are electrocardiographic leads I, II, and V1, and electrogams from the high right atrium (HRA), atrialized right ventricle (ARV), His bundle (HBE), and functional right ventricle (FRV). A represents the atrial electrogram, H the His bundle electrogram, and V the ventricular electrogram.

rhythm with complete right bundle branch block and pulmonary P wave. QRS axis was $+100^\circ$ and non-specific ST-T changes were found in lead III, aVF, and from V1 through V4 (Fig. 2, left panel). The electrocardiogram during the palpitation attack demonstrated a fairly regular rate of 86/min with almost the same QRS configuration as that taken before the attack (Fig. 2, right panel). Furthermore, we could not find an apparent P wave preceding QRS complex in the electrocardiogram during the attack. From these findings, we speculated that the arrhythmia during the attack was ectopic junctional tachycardia.

The tricuspid valve closed 70 msec after the closure of the mitral valve in the echocardiogram, and the septal leaflet of the tricuspid valve was found markedly displaced inferiorly within the right ventricle by two-dimensional echocardiogram. Simultaneous recordings of intracavity potentials and pressures disclosed the existence of the atrialized right ventricle, which showed the right atrial pressure pulse with intracardiac potentials of the right ventricle. Thus, we diagnosed Ebstein's anomaly.

Electrophysiologic study using several intracardiac catheters was done. At the sinus rhythm, PA interval was 20 msec, AH interval 120 msec, and HV interval 40 msec (Fig. 3, upper panel). Wenckebach phenomenon in the AV node appeared at the cycle length of 600 msec in repeat atrial pacing. Effective and functional refractory periods of the AV node were 645 msec and 690 msec respectively. These findings may indicate a deteriorated AV nodal function. Jump up phenomenon in the AV conduction curve was not detected. Furthermore, VA conduction through AV node and concealed by-pass tract was not found by repeat ventricular pacing. Intracardiac electrograms during the attack are shown in the lower panel of Fig. 3. Each V wave was preceded by His bundle depolarization with a HV interval of 40 msec. There was no retro-

Japanese Circulation Journal Vol. 49, April 1985
AV Junctional Tachycardia in Ebstein's Anomaly

Onset of Ectopic Junctional Tachycardia

Fig. 5. Onset of ectopic junctional tachycardia.

The tachycardia is initiated after a single APC (upper panel) or after a single VPC (lower panel).

Shown are atrial premature contraction (APC), ventricular premature contraction (VPC), and P wave in surface electrocardiogram (P). Other abbreviations and symbols are the same as in Fig. 3.

Termination of Ectopic Junctional Tachycardia

Fig. 6. Termination of ectopic junctional tachycardia.

The tachycardia is terminated after timely contraction of the atrium probably due to sinus depolarization (upper panel) or after a single VPC (lower panel).

Abbreviations and symbols are the same as in Figs. 3 and 5.

We evaluated the simultaneous recording of right atrial (RA) pressure pulse and intracavitary electrogram. At the sinus rhythm, "y" wave was slightly elevated, probably due to TR (Fig. 4, upper panel). Once the tachycardia was initiated, however, right atrial pressure increased about twice as much as at the sinus rhythm (so-called "cannon A wave") and then, the patient began to feel palpitation (Fig. 4, lower panel).

As far as we could record, the tachycardia occurred abruptly after a single atrial or ventricular premature contraction (APC or VPC), resembling the form of an escape rhythm (Fig. 5). However, the interval between the ventricular wave of the premature beat and that of the first junctional beat was shorter than that during the sinus rhythm. Also, cycle length of the ventricular depolarizations during the tachycardia was shorter than that during the sinus rhythm. Other electrophysiologic procedures such as repeat atrial and ventricular pacing or atrial and ventricular extrastimulus methods failed to initiate the tachycardia. Similar to the onset, the termination came only after timely depolarization of the atrium or after a single VPC; it was followed by regular sinus rhythm (Fig. 6).

Lastly, we could not find a significant right to left (R-L) shunt by either blood-sampling or dye-dilution methods, and the other catheterization data showed good left ventricular function. We administered a small dose of disopyramide (200 mg/day, 3 × 1, po) to suppress the appearance of APC and/or VPC. As a result, the frequency of the palpitation attack was remarkably decreased without aggravating either the AV nodal or cardiac function. The patient was then discharged and has remained well up to the present time.

DISCUSSION

It is well known that the overall prognosis of

Japanese Circulation Journal Vol. 49, April 1985
Ebstein's anomaly is very poor, and the life expectancy and survival rate are very low. As of now, only 8 patients surviving for 70 years or longer have been reported from the world literature. In Japan, no patient has been reported to have survived into the seventh decade, so the present case is believed to be the oldest patient to our best knowledge. The reason for such long survival may be that the present case is a "Mild Type" according to Takayasu's clinical subdivision of Ebstein's anomaly. That is, the function of his tricuspid valve is fairly preserved, and there is no significant R-L shunt.

Cardiac catheterization study showed that, at the palpitation attack the cannon A wave was formed in the right atrium when the ectopic junctional tachycardia associated with isorhythmic AV dissociation was present. As a result, RA pressure became so high that he began to complain of palpitation.

The junctional pacemaker in the present case had an interesting character. Although this tachycardia appeared most consistent with abnormally enhanced automaticity, it was initiated and terminated abruptly after a single premature contraction. Therefore, this phenomenon cannot be explained solely by the usual ideas of automaticity. On the other hand, electrophysiologic study showed that VA conduction through AV node and concealed bypass tract, the dual AV nodal pathways, and the critical AV conduction delay at the level of AV node and His bundle were all absent. In addition, QRS rate was 80–90/min, and there was AV dissociation during the tachycardia. These findings, then, might exclude reentry utilizing intranodal and extranodal pathways. Previously, Rosen et al applied rules generated by observations of cellular electrophysiologic experiments on delayed afterdepolarization to electrocardiographic intervals in accelerated junctional rhythm by using linear regression and multivariate analysis, and finally they postulated some role of delayed afterdepolarization on the occurrence of accelerated junctional rhythm. Also, it has been shown that the activity of triggered foci can be abruptly initiated or terminated by a single driven premature impulse. In fact, there are several reported cases of ectopic junctional tachycardia which suggest triggered automaticity as the mechanism. Therefore, the occurrence of this tachycardia in the present case might be explained most reasonably by the theory of Rosen et al. And then, it might also resemble the phenomenon of triggered automaticity, although we could not find more direct evidence by electrophysiologic procedures. Moreover, we believe that this is the first case of Ebstein's anomaly to exhibit a recurrent junctional tachycardia which suggests the possibility of triggered automaticity.

Although Kastor et al. previously reported that infranodal rather than intranodal conduction was usually prolonged in patients with Ebstein's anomaly, we found that the AV node was electrophysiologically deteriorated. Lev et al. revealed in their pathological study of Ebstein's anomaly that the AV node was compressed by the enlarged atrial lumen, and that the central fibrous body around the AV junction was abnormally formed, and therefore, not receiving any contribution from the tricuspid valve annulus. Thus, we speculate that the AV junction of the present case must have been damaged morphologically and electrophysiologically through the above process to be led to produce such a specially fashioned automatic focus within it.

REFERENCES

11. HARRIS RHD: Ebstein's anomaly: discovered in a 75-year-old subject in the dissecting laboratory.

Japanese Circulation Journal Vol.49, April 1985

NII-Electronic Library Service
AV Junctional Tachycardia in Ebstein’s Anomaly

Can Med Assoc J 83: 653, 1960


