Unguarded tricuspid orifice diagnosed by echocardiography: a clinical study

Ayhan Kılıç[1], Süheyla Özkutlu[1], Nazlıhan Günal[2]

[1]Cardiology Unit, Department of Pediatrics, Hacettepe University Faculty of Medicine and [2]Department of Cardiology, Social Security Children's Hospital, Ankara, Turkey

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Complete absence of tricuspid valve tissue and apparatus with a normal orifice between the right atrium and the right ventricle is defined as "unguarded tricuspid orifice". Very few case report of this anomaly have appeared in the literature. In this article, we present five cases of unguarded tricuspid orifice, isolated or in combination with other anomalies. All patients were males, aged six days to five years; only one case is alive at present. In our opinion, this anomaly is not so infrequent as it is believed to be, and the diagnosis can be made easily with echocardiography if it is kept in mind.

Key words: unguarded tricuspid orifice, echocardiography.

Complete absence of tricuspid valve tissue and apparatus with a normal orifice between the right atrium and the right ventricle was first described by Kanjuh et al.[1] in 1964 and named "unguarded tricuspid orifice". Very few cases have been reported in the literature since then; the authors are aware of only 10 case reports. In our Department, between December 1995 and February 1998; five patients have been diagnosed by echocardiography to have unguarded tricuspid orifice either as an isolated anomaly or in combination with various other congenital anomalies. The diagnoses were confirmed by catheterization in all five cases. Two of these cases have been published previously[2].

Material and Methods

The patients were all referred to the Department of Pediatric Cardiology with profound cyanosis present from birth and signs and symptoms of heart failure. The patients were all males, ranging in age from six days to five years. After initial routine evaluation with physical examination, telecardiography, electrocardiography and complete blood count, their echocardiographic examinations were made on an emergency basis. Two-dimensional, Doppler and color Doppler echocardiographic examinations were made by the same physician with a Toshiba Sonolayer SSH 160A system. The echocardiographic diagnoses were confirmed and hemodynamic measurements made by cardiac catheterization in all five cases. The diagnoses were further confirmed by necropsy in two cases.

Results

Physical examination was not contributory in any of the patients. Blood counts and electrocardiograms of the patients were nonspecific; telecardiography revealed marked cardiomegaly.

Echocardiography revealed complete absence of tricuspid valvular tissue and apparatus in all five cases (Fig. 1). Right atria and right ventricles were enlarged in all five patients. One patient (Case 5) had a single atrium.

One patient (Case 2), a newborn critically ill at presentation, died during catheterization; the diagnosis was confirmed by necropsy. One patient died awaiting shunt surgery. Three patients (Cases 1, 3 and 4) underwent surgery; two of these died and one (Case 4) is still alive and in good condition with a permanent transvenous pacemaker 18 months after surgery. Diagnoses, management and outcome of the patients are shown in Table I.

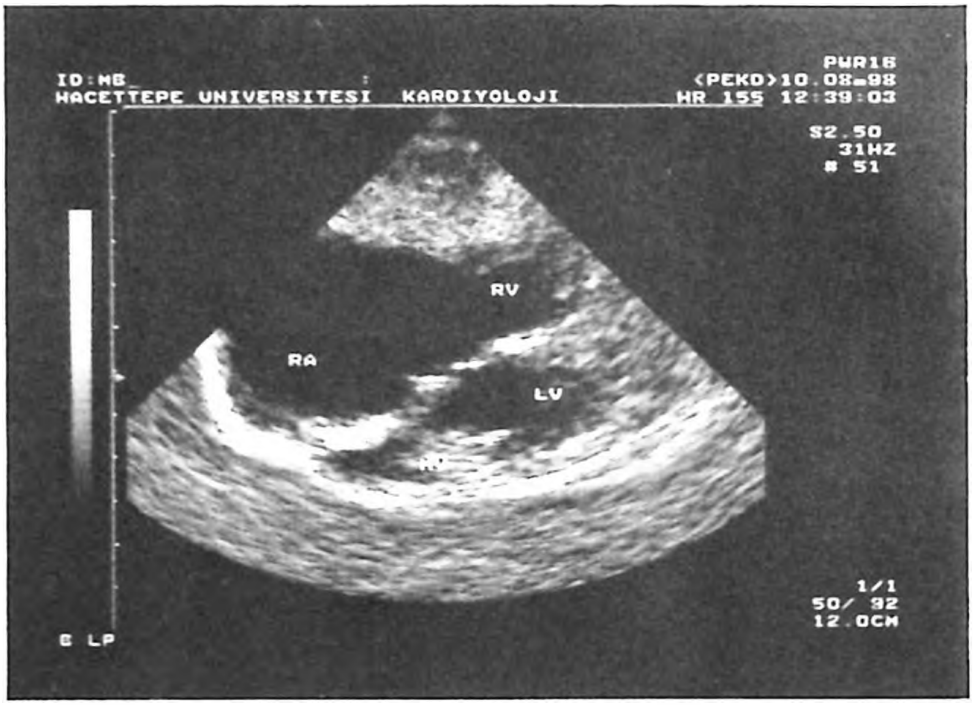

Fig. 1. Echocardiographic view of Case 4 showing complete absence of tricuspid valve tissue and a normal orifice between the right atrium and the right ventricle. RA: right atrium, RV: right ventricle, LV: left ventricle, MV: mitral valve.

Table I. Diagnosis, Management and Outcomes of the Patients

Case	Age	Diagnosis (echocardiography and catheterization)	Management and outcome
1	5 yr	UG	TVR, died postoperatively, necropsy
2	6 d	UG, isolated dextrocardia, pulmonary atresia, PDA, ASD	Died at catheterization, necropsy
3	1 mo	UG, Uhl's anomaly, pulmonary atresia, PDA, dextrocardia	Shunt, died at surgery
4	3 yr	UG, VSD, mild pulmonary stenosis	Alive, ASD and VSD closure, permanent pacemaker implantation
5	2 mo	UG, single atrium, pulmonary atresia, PDA/APCA, hypoplastic left pulmonary artery	Died awating shunt surgery

AG : unguarded tricuspid orifice.
TVR : tricuspid valve replacement.
PDA : patent ductus arteriosus.
ASD : atrial septal defect.
VSD : ventricular septal defect.
APCA : aortopulmonary collateral artery.

Discussion

Complete absence of tricuspid valvular tissue with pulmonary atresia-intact ventricular septum was first observed by Klein in 1938[3]. In this case, necropsy demonstrated almost complete absence of tricuspid valve tissue and rudimentary vestiges of valvular tissue attached to the posterior wall of the right ventricle. Kanjuh et al.[1] reported a second case of pulmonary atresia and intact ventricular septum and complete absence of tricuspid valve, which they termed "unguarded tricuspid orifice". Both of these cases had hypoplastic right ventricles and sinusoidal communications supplying the pulmonary circulation. Gussenhoven[4] reported this anomaly in a neonate with a double-chambered right ventricle.

Anderson et al.[5] reported 46 cases of pulmonary atresia and intact ventricular septum from the Cardiopathological Collection of the Children's Hospital of Pittsburg, of which three cases had unguarded tricuspid orifice and 17 cases had Ebstein's malformation. They observed that all the cases with unguarded tricuspid orifice, but only five of the 17 cases with Ebstein's malformation, had a dilated right ventricular cavity. They pointed out that the two conditions could be differentiated with the examination of the mural leaflet, which is absent when the orifice is unguarded, but displaced in association

with Ebstein's malformation. They suggested the use of cross-sectional echocardiography, especially the subcostal and parasternal long axis approaches, to recognize this feature. In their review, the variant of pulmonary atresia and intact ventricular septum with dilated chambers of the right heart was found to be a lethal combination, which should prompt ventricular decompression as soon as the diagnosis is made.

Magee et al.[6] reported an 11-month-old case of unguarded tricuspid orifice with preexcitation and successful radio frequency ablation of the accessory pathway.

Shahani et al.[7] reported surgical treatment of a seven-year-old girl with an unguarded tricuspid orifice and dilated right heart chambers. They inserted a prosthetic valve at the site of native tricuspid anulus, with prompt improvement in clinical status. After eight years' follow-up, the child is reported to be clinically well and the prosthetic valve functioning normally.

Hornberger et al.[8] and Achiron et al.[9] reported prenatal two-dimensional and Doppler echocardiographic diagnoses of unguarded tricuspid orifice.

In our Department, where approximately 4,500 echocardiographic examinations are made yearly, five cases have been diagnosed to have unguarded tricuspid orifice between December 1995 and February 1998. Although it is not possible with these data to predict the incidence of this rare congenital anomaly, it is the authors' impression that this anomaly is not so infrequent as it is believed to be and that it could be diagnosed easily with cross-sectional echocardiography if it is kept in mind.

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