Koch’s Triangle and the Atrioventricular Node in Ebstein’s Anomaly: Implications for Catheter Ablation

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Introduction and objectives. The development of ablation techniques for supraventricular arrhythmias in patients with Ebstein’s anomaly have led to a need for better understanding of the morphology of the triangle of Koch and the position of the atrioventricular (AV) node in this structure.

Methods. The study involved 17 human hearts: 11 with Ebstein’s anomaly (age range: 37 weeks’ gestation to 1 week after birth) and 6 structurally normal hearts (age range: 35 weeks’ gestation to 2 days after birth). The area of the triangle of Koch was calculated and the length of the AV node and the bundle of His were measured.

Results. The area of the triangle of Koch was significantly smaller in specimens with Ebstein’s anomaly than in control specimens (17.5±4.5 mm² vs. 25.5±6.5 mm²; P<.05). The length of the AV node and its extensions were similar in hearts with Ebstein’s anomaly and normal hearts. The AV node was displaced towards the base of the triangle in 73% of specimens with Ebstein’s anomaly, and the inferior extensions reached the level of the cavotricuspid isthmus. In 91% of specimens with Ebstein’s anomaly, the entry of the His bundle occurred before the apex of the triangle was reached and its length was shorter.

Conclusions. Morphologic findings in this study indicate that performing an ablation procedure close to the base of the triangle of Koch in patients with Ebstein’s anomaly could result in AV nodal block.

Key words: Atrioventricular node. Catheter ablation. Ebstein’s anomaly.

El triángulo de Koch y el nodo AV en la anomalía de Ebstein: implicaciones en la ablación con catéter

Introducción y objetivos. Los avances realizados en los procedimientos de ablación de arritmias supraventriculares en la anomalía de Ebstein (AE) han creado la necesidad de un mejor entendimiento de la morfología del triángulo de Koch (TK) y disposición en dicha estructura del nodo auriculoventricular (AV).

Métodos. Se han estudiado 17 corazones humanos, 11 con AE (intervalo de edades, 37 semanas a 1 semana después de nacer) y 6 estructuralmente normales (intervalo de edades, 35 semanas a 2 días después de nacer). Se calculó el área del TK y se midió la longitud del nodo AV y el haz de His.

Resultados. El área del TK es significativamente más pequeña en los especímenes con AE que en los controles (17,5 ± 4,5 mm² frente a 25,5 ± 6,5 mm²; p < 0,05). En los corazones con AE, el nodo AV y sus extensiones son similares en longitud a los corazones normales. El nodo AV en los especímenes con AE se desplaza hacia la base del triángulo en el 73% y las extensiones inferiores llegan al nivel del istmo cavotricuspídeo (ICT). En el 91% de los especímenes con AE, la entrada del haz de His se produce antes de llegar al ápex del triángulo, y su longitud es más corta.

Conclusiones. Con base en los hallazgos morfológicos obtenidos en este estudio, se puede deducir su utilidad en los procedimientos de ablación en las proximidades de la base del TK por la posibilidad de producir un bloqueo del nodo AV en pacientes con AE.

Palabras clave: Nodo auriculoventricular. Ablación con catéter. Anomalía de Ebstein.

See editorial on pages 633-4
INTRODUCTION

Ebstein’s anomaly (EA) is named after Dr Wilhelm Ebstein, who in 1866 described this congenital heart disease in detail after post-mortem examination of a 19-year-old patient. Few morphological studies have been performed on the atrioventricular (AV) node in childhood and the node may change with age. Previous studies in foetal hearts have shown that the AV node is located within Koch’s triangle (KT) and closer to its base in EA compared with normal hearts. Given the low incidence and wide spectrum of presentation of EA, there is no standardised ablation treatment for cases such as atrial flutter, AV nodal reentrant tachycardia, or accessory AV pathways, all of which have a morphological substrate in the vicinity of KT, with greater ablation complexity in paediatric patients. Therefore, the objective of this study was to analyse the KT area and the conduction system arrangement (AV node and His bundle) in that structure in EA during the perinatal period.

METHODS

Seventeen formalin-preserved perinatal human hearts were studied, 11 with EA and 6 controls. We defined the perinatal period as being from 24 weeks of gestation to 28 days of life. The 11 specimens (6 males and 5 females) with EA belong to the collection of the Paediatrics Service of the Hospital Cruces de Baracaldo. Patients’ ages ranged from 37 weeks to 1 week after birth and the heart weight at time of autopsy was 33.7 (6.4) g (range, 19-42.7 g). The 6 specimens structurally normal hearts were from 4 males and 2 females from 35 weeks of gestation to 2 days after birth. The weight of these hearts was 23.5 (4.5) g (range, 17-35 g). The study was approved by the University of Extremadura Ethics Committee.

For the KT dissection, the right atrium was opened from the tip of the atrial appendage to the AV junction along the right border of the heart. The incision was then extended to the right ventricle and superior vena cava. KT was exposed, inspected, and photographed to document the level of the AV junction. The dimensions of the sides of KT were taken using a ruler and quantitatively verified with calipers (Figure 1). The area was then calculated (area = b×h/2, where b = base; h = height of triangle). The transverse diameter of the coronary sinus orifice (CSO) and the tricuspid annulus were also measured using a string, the length of which was then measured with calipers.

Histological Study

The AV septal junction area was removed in block from each of the hearts. This area contains not only the borders of KT and adjacent parts of the interatrial and interventricular septum but also the superior area of the CSO. The blocks were dehydrated, embedded in paraffin, and serially sectioned at a thickness of 10 µm along an axis perpendicular to the tricuspid annulus. One out of every 10 sections was mounted and stained with Masson trichrome stain and van Gieson stain.

The lengths of the AV node and its extensions, as well as that of the His bundle, were calculated...
Koch’s Triangle in Control Specimens and Those With Ebstein’s Anomaly

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<tr>
<th>Koch’s Triangle</th>
<th>Control</th>
<th>Ebstein’s Anomaly</th>
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<tbody>
<tr>
<td>Base (side b), mm</td>
<td>6.1 (1.5) (range, 4-14)</td>
<td>4.6 (1.3) (range, 2-8)</td>
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<td>Tendon of Todaro (side a), mm</td>
<td>9.5 (2.6) (range, 4-17)</td>
<td>7.8 (1.4) (range, 3-12)</td>
<td></td>
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<td>Septal leaflet of the TV, mm</td>
<td>7.5 (1.5) (range, 5-13)</td>
<td>6.1 (1.3) (range, 4-9)</td>
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<tr>
<td>Area of the triangle, mm²</td>
<td>25.5 (6.5) (range, 16.3-34.5)</td>
<td>17.5 (4.5) (range, 12.3-27.5)</td>
<td>P&lt;.05</td>
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TV indicates tricuspid valve.

The tricuspid valve was malformed to varying degrees in all cases of EA, with significant expansion of the tricuspid valve annulus, 55.1 (10.7) mm (range, 43-75 mm) compared with controls, 28 (4) mm (range, 22-35 mm) (P<.05). The length of the sides and the area of the triangle was smaller in hearts with EA than in control hearts (17.5 [4.5] mm² vs 25.5 [6.5] mm²; P<.05) (Table 1). Furthermore, the transverse diameter of the CSO was significantly higher in specimens with EA than in controls: 3.6 (1.2) mm (range, 1.5-7 mm) vs 2.2 [0.7] mm (range, 1.5-4 mm) (P<.05).

Atrioventricular Node and His Bundle in Control Specimens and Those With Ebstein’s Anomaly

In all of the hearts, the body of the compact AV node was located within KT (Figure 2). In normal specimens, when the CSO was taken as a reference point in the histological sections, only the inferior extensions, and not the body, of the node reached the CSO. However, in most specimens with EA (8 hearts, 73%), given the smaller KT dimensions and greater diameter of the CSO, the body of the AV node ended up located at the level of the triangle base in relation to the CSO (Figure 2).

In control specimens, the body of the AV node had a semi-oval shape and was situated over the central fibrous body (CFB), the muscular AV septum was
formed by the difference between insertion of the tricuspid and mitral rings in the interventricular septum, and the AV nodal artery was situated within the thickness of the node. In specimens with EA, the nodal body had a very irregular shape (semilunar, ovoid, comma, spindle) adapted to KT and the muscular AV septum, which is almost imperceptible in EA as shown in Figure 2B-D, so that the node is arranged almost horizontally on the central fibrous body (CFB) with a distance to the right atrial endocardium varying from 0.1 to 1 mm. In 3 EA specimens (27%), histological sections of the fibromuscular ridge showed the AV nodal artery passing in the thickness of the ridge in the direction of the node (Figure 2).

The body of the AV node extends from the beginning of the bifurcation of the node into its inferior extension to the beginning of the penetrating His bundle in the CFB. We found no significant differences in length of the body of the AV node between controls and specimens with EA. Its length, calculated from the histological sections in control specimens, was 1.5 (0.5) mm (range, 0.8-2.5 mm) and in specimens with EA was 1.7 (0.5) mm (range, 1.1-2.4 mm) (Table 2). The body of the AV node is made up of specialized myocytes (smaller in size than working myocytes), interlinked with one another without a particular orientation, surrounded by a matrix of connective tissue (mainly collagen) surrounds and is between the nodal cells. AVNA indicates AV nodal artery; CFB, central fibrous body; MV, mitral valve; TT, tendon of Todaro; TSL, tricuspid valve septal leaflet.

**Figure 2.** Sagittal histological sections (perpendicular to the tricuspid annulus) stained with Masson trichrome from a control heart (A) and 3 specimens with EA (B, C, and D) at the level of the body of the AV node (delimited with a dashed line red). Note how in the specimens with EA the body is at the level of coronary sinus orifice (CSO) and that the muscular AV septum (muscular area between the insertion of the tricuspid and mitral rings) is not appreciable and has a horizontal orientation. E and F are scanning electron microscopy images of a normal heart (E) and one with Ebstein’s (F). Note the relationship of the AV node with the transitional myocytes and with the working atrial myocytes. The connective tissue matrix (mainly collagen) surrounds and is between the nodal cells. AVNA indicates AV nodal artery; CFB, central fibrous body; MV, mitral valve; TT, tendon of Todaro; TSL, tricuspid valve septal leaflet.

### TABLE 2. Measurements in mm of the Body of the Atrioventricular Node, Inferior Extensions, and His Bundle, in Control Specimens and Those With Ebstein’s Anomaly

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<th>Control</th>
<th>Ebstein’s Anomaly</th>
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<td>Length of the compact body of the AV node, mm</td>
<td>1.6 (0.5) (range, 0.8-2.5)</td>
<td>1.7 (0.5) (range, 1.1-2.3)</td>
<td>≥.05</td>
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<td>Length of the right inferior extension of the AV node, mm</td>
<td>0.5 (0.2) (range, 0.2-1.4)</td>
<td>0.6 (0.1) (range, 0.3-1.5)</td>
<td>≥.05</td>
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<td>Length of the left inferior extension of the AV node, mm</td>
<td>0.4 (0.2) (range, 0-0.9)</td>
<td>0.5 (0.2) (range, 0-0.8)</td>
<td>≥.05</td>
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<tr>
<td>Length of the His bundle, mm</td>
<td>0.7 (0.2) (range, 0.3-1.2)</td>
<td>0.3 (0.2) (range, 0.2-0.7)</td>
<td>&lt;.05</td>
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lower, passing the base of KT to the level of the cavo-tricuspid isthmus (CTI) (Figure 3).

When the body of the AV node is oriented toward the apex of KT and enters the CFB, it is called the penetrating His bundle (Figure 4), which is electrically isolated (by connective tissue) from the rest of the atrial myocardium. The entry point is located in the area of intersection of the anterior and posterior borders of the triangle (membranous septum). In specimens with EA, the anterior border was defined as the fibromuscular ridge or its remnant that marks the true AV junction. Histologically, the beginning of the His bundle was found before reaching the apex in 50% of the control hearts (3 specimens) and in 91% (10 specimens) of those with EA (Figure 4). Furthermore, the His bundle was significantly shorter in length in cases of EA than in controls (Table 2).

**DISCUSSION**

**Main Findings**

Our study shows morphologically and quantitatively that KT is smaller in hearts with EA than in controls. The AV junction at the level of KT is marked by a fibromuscular ridge in the majority of specimens with EA. The body of the AV node and its extensions are shifted toward the base of KT and at the level of the CTI, respectively. Specimens with EA showed abnormal development of the muscular AV septum, since the insertion of the tricuspid and
mitral valve rings are practically at the same height. Histologically, the entry point of the His bundle into the membranous septum occurred before reaching to the apex of KT in our cases with EA (91%, 10 specimens).

Koch's Triangle and the Atrioventricular Node in Specimens With Ebstein's Anomaly. Clinical Implications

Electrophysiologists recognize the potential risk of injuring the conduction system and consequently inducing AV block during catheter ablation in the area of KT and its anatomical surroundings, such as the CTI. The variability in the size and morphology of KT and the presence of the compact AV node and its extensions within its boundaries are essential anatomical determinants during the ablation procedure in paediatric patients.

The KT dimensions obtained in our study in normal hearts in the control group were similar to those previously described by Waki et al.4 in children under 1 year of age. However, KT was found to be smaller in hearts with EA, with 2 important anatomic findings, a larger CSO and the presence of an endocardial fibromuscular ridge marking the anterior boundary of the triangle. This ridge was described in adults by Cabrera et al.7 and later in foetuses by Ho et al.5 The AV nodal artery runs through the thickness of this ridge in 27% of hearts with EA. The topography of the artery requires that the surgeon and/or electrophysiologist have precise knowledge to avoid injury when performing catheter ablation.

The body of the AV node and its extensions in
Our study shows that in hearts with EA in the perinatal period, the inferior extensions, especially on the right (7 specimens, 64%) extend to the base of KT and lie at the level of CTI. It has been suggested that these extensions could provide an anatomical substrate for the so-called “slow pathway” in AV nodal reentrant tachycardia.10

Limitations of the Study

Collecting malformed hearts for histological study is currently difficult, especially when dealing with rare cardiomyopathies such as EA. Therefore, the majority of hearts with EA in this study were 25 or 30 years old. For all these years the hearts have remained fixed in formalin without observing any structural changes to the tissue. In hearts with EA, the morphologic spectrum of dysplasia of the septal leaflet is quite variable and is accompanied by associated cardiac malformations such as atrial septal defect and, at times, pulmonary valve stenosis or patent ductus arteriosus, although there was no mention of arrhythmias in the clinical records of the hearts with EA.

CONCLUSIONS

Knowledge of the dimensions of KT and the orientation of the AV node and its extensions in

Inferior Extensions of the Atrioventricular Node. Clinical Implications

A relatively recent study by Inoue and Becker9 reiterated what Tawara had described in 1906, that is, that the compact AV node has inferior extensions (right and left) and that these extensions, in 44% of adult hearts, may pass the coronary sinus orifice and reach the level of the sub-Eustachian sinus (posterior) of the CTI. A subsequent study2 of structurally normal hearts showed that the extensions increased in length in relation to the age of the individual, but that the inferior extensions did not extend pass the CSO in hearts less than one year of age. Our study shows that in hearts with EA in the perinatal period, the inferior extensions, especially on the right (7 specimens, 64%) extend to the base of KT and lie at the level of CTI. It has been suggested that these extensions could provide an anatomical substrate for the so-called “slow pathway” in AV nodal reentrant tachycardia.10

Penetrating His Bundle: Apex of Koch’s Triangle?

The AV node becomes the penetrating His bundle as we approach the apex of KT. The transition is marked by the entry into the CFB at its junction with the septal leaflet and the tendon of Todaro, both of which, as connective tissue, serve to electrically isolate the His bundle. Anatomically the penetrating His bundle is considered the apex of KT and coincides fluoroscopically with the area in which higher amplitude recordings are obtained on electrogram. However, our histological observations show that the entry point of the His bundle in the fibrous zone is usually prior to the apex of the triangle, especially in specimens with EA. This observation was previously described in some adult hearts, depending on a greater or lesser thickness of the CFB.11 Variability in the position of the His bundle electrogram recording in relation to the KT borders as seen by right atrial angiography has been demonstrated in the electrophysiology laboratory.12 Supporting our findings from control group postmortem hearts, the His bundle electrogram recording is not always at the apex of KT, defined by the anterosuperior border of the tricuspid valve annulus. Furthermore, in hearts with EA, the His bundle is significantly shorter in length than in controls. This could be because EA not only causes an abnormality in the development of the muscular AV septum but also of the CFB and its contents (His bundle). Future studies in the electrophysiology laboratory should correlate our anatomical findings with His bundle electrogram recordings in patients with EA.
EA may be clinically useful with in the setting of surgical treatment of the tricuspid valve and electrophysiological treatment of accessory pathways or other arrhythmic substrates near the base of KT, thus avoiding complications such as complete AV block.

REFERENCES