SECTION ON CARDIOVASCULAR DISEASES

Ebstein’s Malformation of the Tricuspid Valve

Study of a Case Together with Suggestion of a New Form of Surgical Therapy

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Ebstein’s malformation of the tricuspid valve was first described by Wilhelm Ebstein in his publication of 1866. The salient features of this defect which are relatively constant are: (1) A dilated right atrium usually to a severe degree, (2) Downward displacement of the tricuspid valve into the right ventricular portion of the heart, (3) Severe distortion of the posterior and septal leaflets of the tricuspid valve by this downward displacement of their attachments. There may or may not be variable associated defects such as a patent foramen ovale or atrial septal defect, fenestrations of the deformed tricuspid leaflets, diminution in the right ventricular capacity, evidence of tricuspid insufficiency, and conduction defects consisting of right bundle branch block with prolonged PR interval.

This association of congenital defects has been recognized rather infrequently since Ebstein’s description so that in 1937 Yates and Shapiro, reviewing the literature, added the 16th reported case. Kilby and co-workers recently re-reviewed the literature and added five more cases along with one patient surviving closure of the atrial communication.

It is of interest in the light of the frequent association of conduction defects with this malformation that detailed studies by Yates and Shapiro of the right and left bundles of His revealed no abnormalities of the left bundle branch, and although the course of the right bundle branch was somewhat abnormal it conformed in a general way to that of the normal heart.

Thus far medical treatment has consisted of non-specific management of complications as they appear. These have been decompensation (frequently associated with the onset of cyanosis) and arrhythmias. A significant number of these patients die suddenly presumably of ventricular
fibrillation. The surgical approach to this congenital malformation has been limited to attempts at closure of the patent foramen ovale or atrial septal defect if such is present. The goal of this palliative procedure is to eliminate the right to left shunt with its resultant desaturation, hematocrit increase, and paradoxic emboli. However, it is probable that the right to left shunt represents a compensatory phenomenon in these patients and its eradication may hasten death by provoking or aggravating right heart failure. In 1956 the Mayo Clinic group reported one survivor out of five cases operated upon by closure of the atrial defect, the other four died soon after surgery or during induction of anesthesia.3

It is the purpose of this presentation to offer a more definitive surgical procedure based upon the in vivo study of the anatomy in a recently observed case.

Report of Case

The patient was a 10 year old girl who had had cyanosis from birth. Examination and catheterization findings in November of 1952 suggested the diagnosis of Ebstein's malformation. In 1954 the patient was started on digitals as congestive failure was evident for the first time. Also, at that time it was noted that the hematocrit was increasing. By September of 1955 the hematocrit had reached 66 per cent. In November of 1956 she suffered a left cerebrovascular accident which precipitated the decision to seek surgical treatment.

On admission to the University of Minnesota Hospitals for the first time in November 1956, she had right hemiparesis and aphasia. She was deeply cyanotic. Her EKG changes were consistent with the diagnosis of Ebstein's disease with right bundle branch
FIGURE 2: Massive cardiomegaly, much enlarged right atrium, large right ventricle, pulmonary artery segment flattened, decreased pulmonary vascularity.

Figure 4: Shows the triangular piece of atrialized ventricle between the septal leaflet and the true annulus which is to be excluded. The triangular piece of atrialized ventricular tissue between the posterior leaflet and the true annulus has been approximated to the true annulus fibrosis. The triangular area has been excluded by the plicating stitches.

Figure 5: Shows the triangular piece of atrialized ventricle between the posterior leaflet and the true annulus which is to be excluded. The triangular piece of atrialized ventricular tissue between the posterior leaflet and the true annulus fibrosis has been approximated to the true annulus fibrosis.

Figure 6: Shows the triangular piece of atrialized ventricle between the septal leaflet and the true annulus which is to be excluded. The triangular piece of atrialized ventricular tissue between the posterior leaflet and the true annulus fibrosis has been approximated to the true annulus fibrosis.
block, peaked P waves, prolonged PR interval (Figure 1). The roentgen picture was also characteristic (Figure 2). A “box-like” appearance of the heart resulting from the narrowed vascular pedicle and enlarged right atrium were prominent features. A few days after admission, thrombophlebitis of the left leg was noted, and anticoagulant therapy was started. Her condition was considered desperate unless something reparative could be done. It was believed unlikely that simple closure of her patent foramen ovale would provide enough therapeutic benefit to make it worth attempting. The proposed alternative was to utilize total cardio-pulmonary bypass with the pump-oxygenator and under direct vision to attempt a reconstruction of the malformed tricuspid valve leaflets together with closure of the patent foramen or atrial defect. It was thought that this could be done by detaching these malpositioned posterior and septal leaflets and reattaching them more normally. After some slight improvement in her condition, this surgery was planned for December 19, 1956; but she expired during induction of anesthesia.

Examination of the fresh specimen in detail revealed an enormously distended right atrium with a paper-thin wall. The heart, principally the right atrium, contained 3000 cc. of blood. The right atrioventricular annulus measured 19 centimeters in circumference as contrasted to 7 centimeters for the left atrioventricular annulus. The leaflet arrangement of the tricuspid valve was similar to the Ebstein original description (Figure 3). The anterior leaflet was for all intents and purposes quite normal in shape and position though considerably larger than normal. The septal and posterior leaflets were displaced into the right ventricle and were attached far below the normal annular location (Figures 4 and 5). There was no atrial septal defect, but a dilated foramen ovale communication was present.

The Suggested Reparative Procedure

Our previous studies upon this malformation has been based, of necessity, upon the rigid, leathery, formalin fixed specimens available from previous years. These are often completely unsatisfactory for purposes of devising surgical treatment as this case illustrates. In studying possible reparative procedures in this pliable specimen it was noted that the malpositioned leaflets could be repositioned more normally by a simpler maneuver than had been planned.

It was possible, in the fresh specimen, and through a right atrial cardiotomy, to place a row of interrupted silk stitches between the true annulus and the false annulus (i.e. the tissue just above the abnormally placed leaflets) (Figures 3 to 5). When tied, these stitches, having been placed with one bite in the true annulus and the other in the false annulus, pulled the posterior leaflet up to the true annulus by plicating outward a fold of the very thin atrialized ventricular wall (Figure 6). The right coronary artery must be safeguarded in the placement of these stitches. As a result of this plication, the triangular shaped piece of ventricular muscle folded outward would no longer function as an integral part of the right atrium. The identical procedure was carried out for the septal leaflet. This plication maneuver would appear simpler, quicker and more feasible than the previously considered procedure of cutting the leaflets free and reattaching them in a more normal position.

The septal portion of this plication operation would perhaps be somewhat more difficult in that there is not the mobility of the septum comparable to the posterior wall of the right ventricle. Nevertheless, it was possible to oppose the annulus fibrosus at the septum with the septal leaflet. Moreover, it is proposed that any large and obvious fenestrations in the leaflets themselves should be repaired with fine silk sutures and the atrial defect, if present, closed. As the displaced leaflets were brought up to
the annulus in this specimen there was also produced a shortening or snuggling up effect upon the annulus, which in this case seemed beneficial. If considerable tricuspid incompetency were still present after plication, the diameter of the atrioventricular ring could be further shortened by placement of a few horizontal mattress stitches tied over small Ivalon pledgets, such as has been utilized for the correction of mitral insufficiency.

One is of course disturbed by the paucity of valve tissue present in the small nubbin-like septal leaflet, but it seemed quite likely that the other two leaflets with their more abundant leaflet tissue should be adequate to complete the atrioventricular closure during ventricular systole, particularly since the anterior leaflet in this specimen had undergone compensatory enlargement. In summary, the multiple potential accomplishments of this plication operation proposed for Ebstein's malformation would be neutralization of the contrary acting ventricular tissue in the atrium, the reduction in annular circumference, and repositioning of the malposed tricuspid leaflets so that they could function more effectively.

Discussion

The right side of the Ebstein heart beats inefficiently and often paradoxically and this surgical procedure should correct these abnormalities. That is, as the right atrium of the Ebstein heart contracts in systole, the ventricular portion which has been atrialized, relaxes in diastole, and when the right atrium is in diastole, the atrialized ventricular portion is contracting. Since these two triangular shaped areas of the atrialized ventricle would be excluded by the plication with stitches as described, the Ebstein heart might be freed of this paradox and recover some efficiency on this basis alone.

According to Kilby's recent study, approximately one-third of the recorded deaths from Ebstein's malformation were the result of congestive heart failure. Thus, it seems likely that a majority of these patients have congestive heart failure in the terminal stages of their disease. In many Ebstein patients there is no disagreement over the presence of clinically obvious tricuspid insufficiency. However, in other patients this incompetency is not immediately obvious probably being masked by the greatly enlarged and distensible right atrium allowing considerable regurgitation of blood by serving as a reservoir and thus acting as a buffer for the systemic veins and the liver. Also, there is the decompressing effect of a patent foramen ovale which may postpone clinically obvious right heart failure. The proneness of these patients to fatal arrhythmias is more difficult to explain, but it is clear that this complication plays a consistently more important role than in most other forms of congenital heart disease. Moreover, cardiac catheterization in this malformation has been more hazardous because of this danger of a serious arrhythmia. It is hoped that the proposed surgical procedure by establishment of more effective right heart function with consequent reduction in the atrial over-distention present might lessen their susceptibility to these arrhythmias. Lev et al have contributed additional information upon these arrhythmias.
exhaustively studying a case of Wolf-Parkinson-White Syndrome (short
PR interval with a prolonged QRS) in an Ebstein heart. These investi-
gators found an anatomic basis for the arrhythmias occurring in their
patient. These abnormalities consisted of a right atrioventricular com-
munication outside the usual conduction system, a communication of the
right bundle branch and the right side of the septum, and encasement of
the right branch in dense fibroelastic tissue. One gains the impression
from their study that these extra-conduction pathways consisting of an
intermediary muscle bundle with numerous fasiculi to the right atrial
appendage and the parietal wall of the right ventricle are the most im-
portant elements in the genesis of these conduction irregularities. This
may account for the arrhythmias found in the Ebstein heart and also for
the unexplained deaths due to this disease. If so, it is possible that the
suggested operative technique might alter beneficially this irregular path-
way of conduction by deliberately placing the plicating stitches close
enough together so as to interrupt these abnormal atrioventricular path-
ways.

The proposed operation would necessitate total cardiopulmonary bypass
with the pump-oxygenator as previously described.\textsuperscript{5, 6, 7} Obviously, a trial
in patients who have not reached the terminal stages of myocardial de-
compensation would offer the best test of this hypothesis.

SUMMARY

A case of Ebstein's malformation of the tricuspid valve has been studied.
Because of the relative rarity of this defect and the lack of any adequate
medical or surgical treatment, a suggested operation is presented for
direct attack upon the deformities. Under direct vision by use of total
cardiopulmonary bypass the paradoxically contracting atrialized ventric-
ular tissue would be excluded by plication. At the same time, tricuspid
valvular function would be improved by the bringing up of the abnormally
placed tricuspid leaflets to their true annulus. Also, any patent foramen
ovale or atrial septal defect found present would be closed, and fenestra-
tions in the leaflets of the tricuspid valve repaired by fine stitches. The
goal of this proposed operation would be the creation of more effective
right heart function.

RESUMEN

Se presenta un caso de malformación de Ebstein de la válvula tricúspide.
Debido a la falta de tratamiento médico o quirúrgico adecuado, proponemos
una operación para la corrección de esta anormalidad. La operación tiene por
objeto eliminar funcionalmente el tejido ventricular situado por arriba de
das valvas anormalmente desplazadas de tricúspide, mediante la fijación
de su borde de inserción al anillo auriculo-ventricular, utilizando un método
cielo abierto y bajo visión directa con ayuda de circulación extracorporea
ey exclusión cardio-pulmonar circulatoria temporal. Se piensa que la función
de la tricúspide mejoraría de esta manera. Simultáneamente una comuni-
cación interauricular o un orificio de Botal persistente, así como posibles
fenestraciones en las valvas de la tricúspide, podrían ser reparados ade-
cuadamente. La meta de la operación sería la creación de un corazón derecho funcionalmente más efectivo.

RESUME
Un cas de malformation d'Ebstein de la Valvule tricuspid a été étudié. A cause de la rareté relative de cette lésion et de l'absence de traitement médical ou chirurgical adéquat, nous avons suggéré une technique corrective. La portion "auricularisée" du ventricule, avec contraction paroxysmale, serait exclue vers l'extérieur par plicate, sous vision directe à l'aide de l'appareil coeur-poumon. La fonction de la tricuspid étant améliorée du même coup par l'élévation des feuillots valvulaires au niveau de l'anneau trio-ventriculaire. Les anomalies associées telles que "foramen ovale" ou communication interauriculaire seraient suturées et les orifices dans les feuillots tricuspidiens pourraient être réparés en même temps.

Le but de cette intervention est l'amélioration de la fonction ventriculaire droite.

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REFERENCES