Late Dysrhythmias and Disturbances of Conduction following Mustard Operation for Complete Transposition of the Great Arteries

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SUMMARY Between 1964 and 1971, 49 patients were discharged from hospital following atrial baffle repair for transposition of the great arteries. Electrocardiograms of 47 of these patients were available for review seven months to eight and a half years (average 31 months) after operation. Of the 46 patients in sinus rhythm prior to operation, 40 (87%) were in sinus rhythm with normal atrioventricular conduction when last seen. Four patients (8.7%) had benign supraventricular dysrhythmias: three were junctional, one was atrial. None was noted to experience rapid dysrhythmias. Two patients showed evidence of abnormal atrioventricular conduction.

IN PATIENTS with complete transposition of the great arteries the Mustard operation has become an established procedure to correct the abnormal circulatory pathway. However, recent reports have focused attention on dysrhythmias occurring in these patients after operation and modifications in surgical technique have been suggested to lower their incidence. We have noted fewer rhythm disturbances late postoperatively than others. This investigation was undertaken to assess the incidence and type of dysrhythmias present at the time of hospital dismissal and on subsequent review, and to determine how late dysrhythmias and disturbances of conduction might be related to surgical technique.

Material and Methods

Between 1964 and 1971, 49 patients were discharged from Green Lane Hospital after a Mustard type of atrial baffle repair for complete transposition of the great arteries. In 47 of these patients, aged 11 days to 58 months at the time of operation (fig. 1), electrocardiographic data obtained seven months to eight and a half years (average 31 months) postoperatively (fig. 2) were available for review and comparison with earlier records. Three of the 47 patients later died, two from noncardiac causes and one from pulmonary venous obstruction.

Follow-up studies were not obtained in two patients, each of whom was in sinus rhythm at the time of hospital discharge. One died with pulmonary venous obstruction and one died suddenly nine months after operation.

Resting electrocardiograms were analyzed to determine the anatomical origin of the activation wave and its discharge and conduction sequence. Patients were considered to be in sinus rhythm postoperatively when similar P waves preceded each QRS complex, the P-R interval was constant and the frontal plane P wave axis was similar to that seen before operation.

Surgical Features

All but one of the 47 patients received prior palliation either by surgical atrial septectomy (22) or atrial septostomy (24). Septectomy consisted of excision of the valve of the fossa ovalis and vertical incision of the limbus during a short period of circulatory arrest under either mild hypothermia or normothermia, except in one instance where a Blalock-Hanlon procedure had been performed elsewhere.

The atrial baffle consisted of pericardium in all instances, but the technique of repair varied in several important respects relative to rhythm disturbances.

1) In 22 patients, aged 21 to 58 months (fig. 1), conventional cardiopulmonary bypass was used with mild hypothermia to 30°C. Twenty of this group had had an earlier atrial septectomy, and one a Blalock-Hanlon operation. The two caval venous return cannulae were inserted through separate purse string sutures placed in the lateral right atrial wall adjacent to the origins of the cavae. In a few of the earliest patients operated upon, the atrial wall purse string was first excluded in a curved clamp, but in most, to minimize damage to the sinus node area, a clamp was not used. The right atrium was opened with a vertical incision extending between the caval cannulae, which followed the line of the previous atriotomy used for atrial septectomy and lay in front of the crista terminalis.

2) In 25 patients, aged 11 days to 36 months, profound hypothermia with circulatory arrest and limited cardiopulmonary bypass was used. Two of this group had had an earlier atrial septectomy and 23 balloon atrial septostomy. With the profound hypothermia technique only one venous return cannula was required and this was inserted through the tip of the right atrial appendage. In this group, the right atrium was opened through either a vertical incision in front of the crista terminalis identical to that just described or, in the last 15 infants, with an oblique incision commencing just below the atrial appendage purse string and passing either directly backwards toward, but not into, the crista ter-

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minalis, or downwards toward the inferior vena cava.

3) The extent to which the atrial septum was excised at the time of the atrial baffle operation depended on the size of the atrial septal defect. Following septectomy, which included division of the limbus, a large defect was always present and at the time of baffle repair further excision was required mainly in front of the right pulmonary veins. Following septostomy, the limbus remained intact and excision always involved this area in addition to the remnants of septum anterior to the entry of the right pulmonary veins. These maneuvers always destroyed the middle internodal tract and may have damaged the anterior and posterior tracts (fig. 3 left). In later infants only the right half of the limbus was excised (directly behind the superior vena caval orifice) in the hope of preserving the anterior internodal tract (fig. 3 left).

4) The atrial baffle suture line consisted of continuous sutures and passed 10-15 mm in front of the superior vena caval orifice to avoid the sinus node. Its siting varied, however, in relation to the coronary sinus ostia and atrioventricular node (fig. 3 right). Thus, in the first seven patients (all operated upon using heart/lung bypass) the suture line passed onto the tricuspid ring, or base of the septal tricuspid leaflet, in front of the atrioventricular node and coronary sinus ostium. In the remaining 39 patients the coronary sinus ostium was first cut downwards into the left atrium, dividing the posterior internodal tract, and the suture line was placed across the floor of the opened coronary sinus behind the atrioventricular node. With both techniques coronary sinus drainage entered the caval compartment behind the baffle and the baffle suture line crossed the posterior internodal tract at two or more points and the anterior tract once (fig. 3 right).

5) At the time of atrial baffle repair five of the 47 patients had surgical closure of a moderate or large ventricular septal defect through a right ventriculotomy. In two of these and in an additional patient with an intact ventricular septum, left ventricular outflow obstruction was also relieved.

### Postoperative Dysrhythmias and P Wave Changes

#### Early Postoperative Dysrhythmias

Rhythm disturbances were common in the first postoperative week. Paroxysmal supraventricular tachyarrhythmias occurred in three patients. One settled spontaneously, another responded to digitalization and one required electrical cardioversion. The commonly encountered dysrhythmias were 'passive,' i.e., there was depression of the sinus pacemaker (or sinoatrial block) with atrial or junctional escape rhythms, sometimes with atrioventricular dissociation. These dysrhythmias produced no important morbidity and resulted in no deaths. They occurred in 23 patients and are not analyzed further in this report.

#### Incidence of Late Postoperative Dysrhythmias

The incidence of late rhythm disturbance has been related to the year of review in figure 2 and to the rhythm present preoperatively and at dismissal from hospital in figure 4.

1) One patient was in junctional rhythm preoperatively and remained in this rhythm 48 months after operation. Following exercise, junctional rhythm persisted and the ventricular rate increased from 56 to 72 beats per minute.

2) Thirty-nine of 46 patients in sinus rhythm prior to atrial baffle insertion (fig. 4) were in sinus rhythm at discharge and 36 of these have remained in this rhythm with normal atrioventricular conduction subsequently, the time of the last review varying from seven months to eight and a half years (average 29 months). Two of these patients died eight and eleven months after operation. Both were in sinus rhythm a month earlier and their deaths were not due to dysrhythmias (subdural hemorrhage and pulmonary venous obstruction, respectively).

Three of the 39 patients in sinus rhythm at discharge have subsequently shown disturbances of rhythm or conduction.
In one, periods of slow atrial dysrhythmia alternated with a faster sinus rhythm during the recording of a standard 13 lead electrocardiogram 26 months postoperatively (fig. 5). Another was noted to have junctional rhythm 15 months after reoperation for narrowing of the superior vena caval compartment two years after atrial baffle repair. Conducted sinus beats were seen at a faster heart rate following exercise. In the third patient, who had an intact ventricular septum, complete heart block was present during the early postoperative period but sinus rhythm with normal conduction appeared on the fifteenth day after operation and persisted until his discharge ten days later. Four months after operation he was again found to have complete heart block which was still present 13 months later. He remains asymptomatic on long acting isoproterenol with a narrow QRS complex (0.07 sec) and a ventricular rate of 52/min.

(3) Five of the 46 patients in sinus rhythm preoperatively had junctional rhythm at discharge. Three of these did show some sinus activity at this time and were found to be in sinus rhythm seven, 13 and 25 months postoperatively. Two of the five were still in junctional rhythm at 43 and 53 months although the latter patient was in sinus rhythm when reviewed at 26 months. Both reverted to sinus rhythm after exercise but with occasional sinoatrial block (fig. 6).

(4) Two of the 46 patients in sinus rhythm preoperatively had a passive atrial dysrhythmia at dismissal. One was in sinus rhythm when seen 35 months postoperatively. The other continued to show a passive atrial dysrhythmia up to 53 months, but at 64 and 77 months postoperatively there was sinus rhythm with first degree heart block. On the latter occasion a more advanced degree of atrioventricular block developed after exercise (fig. 7).

In summary, of the 46 patients in sinus rhythm before operation 40 (87%) were in sinus rhythm with normal atrioventricular conduction when last reviewed; four (8.7%) had a benign supraventricular dysrhythmia (three junctional, one atrial) but in one of these this followed a second operation; two had an intact sinus mechanism but there were disturbances of atrioventricular conduction.

<table>
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<th>PRE-OP</th>
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<td>1 junctional</td>
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FIGURE 3. Left: Diagram of opened right atrium and tricuspid orifice to show the position of the three internodal pathways. There is an atrial septal defect produced by balloon septostomy. The interrupted lines show the position of the incisions for excision of the atrial septal remnant and for division of the coronary sinus opening downward into the left atrium. Right: The atrial septum has been excised and the coronary sinus laid open. The dotted line follows the position of the atrial baffle suture line when it was deviated anterior to the coronary sinus and the atrioventricular node in the first seven patients operated upon. The points of complete interruption of the middle and posterior internodal tracts are clearly displayed and also the relationship of the baffle suture line (interrupted line) to the internodal tracts.

FIGURE 4. Relationship of late postoperative electrocardiographic findings to rhythm present prior to operation and at hospital dismissal.
Relation Between Dysrhythmia and Duration of Follow-up

While the data presented in figure 2 might suggest that the incidence of dysrhythmia increased with the duration of follow-up, closer examination does not confirm this impression. Thus in two of the three patients with junctional rhythm in the fourth and fifth years postoperatively this was present at the time of hospital discharge and in the remaining patient it appeared following reoperation.

Relation Between Arrhythmia and Surgical Technique (Table 1)

(1) In the first seven patients in this series the baffle suture line passed anterior to the coronary sinus ostium and atrioventricular node. These patients were operated upon using cardiopulmonary bypass which required double caval cannulation. On last review at an average of 52 months postoperatively, five patients were in sinus rhythm, one had first degree heart block and one junctional rhythm.

(2) In the remaining 39 patients in sinus rhythm preoperatively the coronary sinus opening was cut downwards into the left atrium and the baffle suture line was positioned across the floor of the opened coronary sinus posterior to the atrioventricular node.

Fourteen of this group were corrected using cardiopulmonary bypass with double caval cannulation. On final review at an average of 30 months postoperatively, one patient had complete heart block and two supraventricular rhythm disturbances (one junctional, one atrial). The junctional rhythm, however, appeared only following reoperation which required repeat double caval cannulation.

Twenty-five of this group were corrected using a single right atrial cannula in association with circulatory arrest and profound hypothermia. All were in sinus rhythm preoperatively and at hospital dismissal and only one had an atrial dysrhythmia at final review which averaged 25 months postoperatively. Twenty-three of the 25 patients were less than two years of age at the time of operation, 15 of these being less than one year (fig. 1).

Comparing double caval cannulation using cardiopulmonary bypass with profound hypothermia (table 1) the incidence of supraventricular dysrhythmias in patients in sinus rhythm before operation with the former technique was three in 21 (14%) and with the latter, one in 25 (4%). The numbers are too few to permit statistical conclusions. In the hypothermia patients the majority (23) had preliminary balloon septostomy while in the bypass patients the majority (20) had surgical septectomy. The length of follow-up was shorter in the hypothermia group.

Other variations in surgical technique had no apparent effect on the incidence of dysrhythmias. Thus, the siting of the right atriotomy was unrelated as vertical, transverse and oblique incisions were all used in the hypothermia group. The same was probably true for the extent of excision of the portion of atrial septum superior to the limbus as preservation of its anterior half (including the anterior internodal tract) was also introduced halfway through the hypothermia series. All patients undergoing ventricular septal defect repair or relief of pulmonary stenosis remained in sinus rhythm with normal atrioventricular conduction.

Postoperative P Wave Changes

The mean frontal plane P wave axis ranged from +30° to +90° postoperatively and was not significantly different from the preoperative axis. There was, however, a striking alteration in P wave amplitude after operation best seen in lead two (fig. 8). In most patients the P waves in standard leads were of low voltage postoperatively, and frequently bifid in shape. In 15 of the 18 where the P wave measured more than 2.5 mm preoperatively it was decreased by 2 mm or more.

Discussion

This report is concerned with dysrhythmias present at the time of hospital discharge three to four weeks after atrial
baffle repair and on later review. Although early postoperative dysrhythmias were commonly encountered they rarely caused significant morbidity. Previous publications have reported a high incidence of late dysrhythmias but 87% of our patients appeared to be in sinus rhythm with normal atrioventricular conduction at last review. The report of El-Said et al., which is comparable to ours as it reviews 60 surviving patients also operated upon between 1964 and 1971, not only showed an incidence of late dysrhythmia in the region of 90% but also an increase in the incidence of rapid dysrhythmias as the length of follow-up increased, and a late mortality from this cause. No patient in the present series had a rapid (or active) dysrhythmia after hospital discharge and late death from this cause has not been documented although the death of one of the two patients in whom follow-up studies were not available could have been due to an abnormal rhythm. The incidence of late 'passive' dysrhythmias was low (8.7%) and as in El-Said's series did not increase progressively. Only one appeared spontaneously for the first time after hospital dismissal. It was more common for a benign dysrhythmia present at dismissal to disappear subsequently. Atrioventricular block has also been infrequent, one patient developing first degree heart block and one complete heart block. Admittedly, prolonged monitoring, during sleep, may have disclosed a higher incidence of dysrhythmias but there is no evidence to date that this is an important factor from the clinical standpoint.

As all the operations in this series were performed by, or under the supervision of, one surgeon the technique has been consistent and changes have been accurately documented. As a result, reliable information is available concerning several matters which remain controversial.

(1) Division of the free wall of the coronary sinus ostium in a vertical direction downward into the left atrium (which divides the posterior internodal tract) is not a cause of late dysrhythmia. This statement is substantiated by the knowledge that the 25 patients in whom this maneuver was performed using the profound hypothermia technique had sinus rhythm at dismissal and 24 remained in sinus rhythm subsequently. In addition, as the baffle suture line passed posterior to the atrioventricular node across the floor of the opened coronary sinus in these patients, it follows that this is also unrelated to late dysrhythmias. Consequently, because this technique also widens the caval compartment beneath the baffle and drains coronary sinus blood into the caval compartment it remains our method of choice.

(2) Claims that placement of the baffle suture line in front of the atrioventricular node will reduce dysrhythmias are not supported by this series. Our only example of first degree heart block occurred with this technique. Moreover we are concerned that when sutured in this position later changes in the baffle may distort the tricuspid leaflet and precipitate tricuspid incompetence.

(3) The results suggest an increased incidence of dysrhythmias when two venous cannulae are inserted into the lateral right atrium adjacent to the origins of the cavae, when compared with cannulation of the right atrial appendage alone. This difference may be related to damage to the sinus node itself or the sinus node artery during placement of the superior caval cannula, particularly when the artery runs an aberrant course lateral to the superior vena cava before reaching the node. Rodriguez-Fernandez et al. have produced data suggesting that late dysrhythmia is related to sinus node damage rather than division of internodal conduction pathways in the right atrium. Our findings and the electrophysiological studies of Gillette et al. would support this. Whether the late changes in the pericardial baffle may damage the node remains speculative: in this series the suture line has been placed 10 mm or more in front of the node to lessen this possibility. Clearly clamps should not be applied to the right atrium for placement of the superior vena caval cannula nor should the atria be pierced for pressure monitoring medial to the superior vena cava for the artery normally courses in this area.

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<thead>
<tr>
<th>Suturing anterior to coronary sinus</th>
<th>Bypass</th>
<th>Technique</th>
<th>Hypothermia</th>
<th>Rhythm</th>
<th>Disturbance of Conduction</th>
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<tr>
<td>Suturing posterior to coronary sinus</td>
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<td>25</td>
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FIGURE 7. *Top* — sinus rhythm with first degree heart block (P-R 0.26 sec). *Bottom* — Following exercise — second degree heart block (Wenckebach).
Other technical factors seemed less important. It appears unlikely that late dysrhythmia is more common when the anterior tract is destroyed or damaged, in addition to the posterior and middle tracts. Currently, however, in an attempt to preserve the anterior tract (which is probably the most important of the three) the anterior half of the limbus and adjacent upper atrial septum are preserved and the baffle suture line in this region picks up the free edge of the limbus before crossing medial to the superior vena caval opening.

The changes in P wave amplitude and shape after atrial baffle repair raise the possibility that what has been called sinus rhythm may have a different origin and conduction sequence from normal. The study of Sealy et al. in dogs suggests that after exclusion of the sinus node from the rest of the atrium the “sinus” rhythm which becomes re-established in about two weeks originates in the coronary sinus area. It is possible that the early postoperative dysrhythmias seen in patients are related to division of the internodal pathways, as seen in the animal experiments. El-Said et al. postulated silent retrograde conduction from an ectopic focus to explain ‘sinus-like’ P waves in patients after operation. They later produced evidence that the ectopic focus could be in the region of the atrioventricular node. Preservation of this region may allow retrograde conduction to the left atrium or along the anterior internodal tract with subsequent activation of the right atrium in an approximately normal direction. In this case the technique of cutting back the coronary sinus could be advantageous as it keeps the posterior baffle suture line well clear of the atrioventricular node and conduction tissues around it. Whether “sinus” rhythm after Mustard atrial baffle repair results from this mechanism or truly originates from the sinus node remains speculative. In either case the rhythm appears acceptable and certainly accumulated surgical experience suggests that every precaution should be taken to preserve the sinoatrial node and its arterial supply. If possible at least one of the internodal tracts should be preserved but our experience suggests this is a secondary consideration. Adequate caval channels, unpaired tricuspid valve function and above all unobstructed pulmonary venous return should have precedence, and are of critical importance in the smaller infant.

References
Late dysrhythmias and disturbances of conduction following Mustard operation for complete transposition of the great arteries.
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