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## Cardiac arrhythmias after atrial surgery in children

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## SUMMARY

Thanks to the rapid development of cardiac surgery, the quality of life and the life expectancy of children with congenital heart disease has improved substantially during the last decades. However, one of the risks of cardiac surgery is that the conduction system of the heart, which is almost invisible for the surgeon, is damaged. This thesis deals with the cardiac arrhythmias seen after two types of operation in the atrium of the heart; the Mustard operation for transposition of the great arteries and the surgical repair of the atrial septal defect of the secundum type. The motive for the studies collected in this thesis was the fact that we saw a high incidence of arrhythmias after both operations, requiring drug treatment or pacemaker implantation, and occasionally resulting in sudden unexpected death. The objectives of our work were to determine the incidence, the severity and the natural history of the arrhythmias seen after these types of atrial surgery. An even more important goal was to detect the causes of these arrhythmias, and to find the starting-points for elimination of them. Chapter 1 gives the information essential for an understanding of the problem; information about the anatomy of the conduction system, the technique of the different operations, as well as the clinical consequences of injury of the conduction system.

In the study described in Chapter 2, we analyzed the arrhythmias seen after long-term follow-up in 50 children after the Mustard operation, and in 204 children after repair of an atrial septal defect. We found a 42% incidence of arrhythmias many years after the Mustard operation. The incidence of arrhythmias was influenced unfavorably by the cardio-pulmonary bypass time, and favorably by the use of cardioplegia and by selective cannulation of the superior vena cava. In patients with an atrial septal defect, the incidence of arrhythmias was less, but still 10% of the patients had severe arrhythmias, related to the type of atrial septal defect and the presence of partial abnormal venous drainage. The number of children with arrhythmias increased significantly, if cardio-pulmonary bypass was used instead of hypothermia. We wondered if the cannulation could have damaged the sinus node area directly or indirectly and thus causing arrhythmias. The incidence of sudden death was high in children after the Mustard operation and was significantly related to the presence of atrial flutter.

In 1974 the section of pediatric cardiology, Houston, USA, published their improved results after surgical modifications of the Mustard operation. Chapter 3 describes the longer follow-up of the patients operated upon in Houston with these modifications. Although the early results were favorable, long-term follow-up showed a progressive loss of sinus rhythm (50% after 5-8 years) and a gradual development of active and passive arrhythmias.

Although sudden death, reoperations and pacemaker implantations overshadow the results of the Mustard operation, most children are asymptomatic and do remarkably well. We studied 36 of these "asymptomatic" children hemodynamically and electrophysiologically,  $7.7 \pm 2.5$  years after the operation and analyzed their electrocardiograms and 24-hour Holter recordings (Chapter 4). A surprising number of hemodynamic problems was found, such as systemic- and pulmonary venous baffle obstructions, baffle leakage and left ventricular outflow tract obstruction. Only 31% had no hemodynamic abnormalities. Moreover, the electrocardiogram showed sinus rhythm in only 42%, and only 6% of the children showed no abnormalities on their 24-hour Holter recording. Electrophysiologic studies revealed often severe sinus node dysfunction in 84%, and electrophysiologic abnormalities of the atrial myocardium in 63% of the children. In 32% of the children, supraventricular tachycardia was induced during the electrophysiologic study. The atrioventricular node function was normal in most children. There was no positive influence of the surgical modifications applied over the years.

In our general discussion (Chapter 7) we state that the sinus node area and the atrial myocardium are damaged by the Mustard operation, causing arrhythmias and sudden unexpected death by tachy-arrhythmias. This injury could not be prevented by several surgical modifications and seems to be inherent to the technique of the Mustard operation. Therefore, we advise to replace the Mustard operation by the arterial switch operation (Chapter 1), although the long-term results of this technique are not yet fully known.

In order to understand the relatively high incidence of arrhythmias after repair of an atrial septal defect, we undertook two further studies. First we performed an electrophysiologic study in 40 children with atrial septal defect preoperatively (Chapter 5). We found an abnormal electrophysiologic function of the sinus node in 83% of the children. The atrial myocardium also behaved abnormally. The electrophysiologic abnormalities of the atrioventricular node were less important. The electrocardiogram of 15% of the children showed first-degree atrioventricular block and 35% of the children had prolonged periods of accelerated atrial rhythm on their 24-hour Holter recording. Both abnormalities were significantly less frequent after repair of the atrial septal defect. It seemed reasonable that these electrophysiologic abnormalities and arrhythmias were the result of stretch of the right atrial wall and the adjacent conduction tissue by the prolonged volume load of the atrial septal defect. However, we could not find a correlation with age or shunt size in our group. This could be influenced by the fact that we had to exclude the data of 11 young patients, whose sinus node measurements appeared to be influenced by premedication with thiopental.

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Analysis of the 24-hour Holter recordings of 46 children before, and one year after, atrial septal defect operation (Chapter 6) showed that arrhythmias seen after operation partially consist of arrhythmias already present before operation and partially of arrhythmias acquired as a result of the operation. The arrhythmias present before operation tended to disappear in the years after repair of the atrial septal defect. This is not seen after operation of an atrial septal defect in adults.

The results of our former retrospective study (Chapter 2) showed that cannulation could possibly be responsible for the injury of the sinus node area. Therefore, our second study in patients with an atrial septal defect (Chapter 6) was a prospective study in 50 children, who were operated upon with a change in cannulation technique. The superior vena cava was cannulated directly instead of through the right atrial appendage. The incidence of post-operative arrhythmias decreased significantly from 23 to 10%. Moreover, no severe arrhythmias (e.g. atrial flutter) were seen during long-term follow-up of the prospective group, compared with 10% in the retrospective group.

In our general discussion (Chapter 7) we state that arrhythmias after surgical repair of atrial septal defect partially consist of arrhythmias present before operation and partially of arrhythmias caused by the operation. Early repair of atrial septal defect probably will increase the chance that the electrophysiologic abnormalities and arrhythmias, present before operation, will disappear after operation. Moreover, the development of arrhythmias after surgical repair of an atrial septal defect can be reduced significantly by surgical modifications, such as selective cannulation of the superior vena cava. We suggest that this cannulation technique be used in all types of cardiac surgery.

These studies show that early prospective studies should be done after every new type of cardiac surgery to analyze the causal factors of arrhythmias and to find methods of prevention.