

Arrhythmias in patients with surgically treated atrial septal defects

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Summary

Atrial arrhythmias complicate the clinical course of adult patients with an atrial septal defect. Atrial flutter is more prevalent in younger patients, and frequently regresses after surgical defect closure. Atrial fibrillation however, which results from a chronic underlying disease, rarely reverts to sinus rhythm after surgical repair. While younger patients with atrial flutter clearly benefit from de-

fect closure alone, this is not evident in older patients with fibrillation, who may need an additional anti-arrhythmic procedure concomitant to defect closure.

Key words: arrhythmia; CHD; septal defects; prognosis

Introduction

An atrial septal defect (ASD) is the most common congenital heart defect detected in adulthood, as it rarely causes symptoms in childhood [1]. The main reasons why patients with an ASD develop symptoms such as reduced exercise tolerance and stroke, are right ventricular dilatation and dysfunction [2], pulmonary hypertension secondary to increased pulmonary vascular resistance [3], and arrhythmias [4]. Of the latter, atrial flutter and atrial fibrillation (AF) are by far the most common encountered in the adult population. They cause important morbidity, mainly related to the occur-

rence of transient ischemic attacks and stroke [2]. While atrial flutter and AF frequently occur in adults before closure of an ASD, they may also persist or occur after the defect has been closed [4]. In the past, doubt has been expressed whether surgery really improves long-term outcome [6]. Some recent studies suggest that defect closure does not alter the occurrence of atrial flutter/fibrillation [5]. This review addresses the effect of closure of an ASD on the incidence of atrial arrhythmias and/or cerebrovascular accidents by comparing studies of others [4, 8] and our own [7].

Impact of surgical atrial septal defect closure on arrhythmia

There are three major clinical publications addressing the question of the effect of surgery on the incidence of atrial flutter/fibrillation in adults with and ASD [4, 7, 8]. Together these studies looked at 616 adults (213 in Toronto, 211 in Berlin, and 192 in Madrid) who underwent surgical closure of an ASD within the oval fossa. The methods and results of the Toronto and Berlin groups, which were conducted independently, are remarkably similar. They examined the incidence of atrial flutter

and/or fibrillation in adults, before and after surgical closure of an ASD with significant left-to-right shunting, defined as a pulmonary (Qp) to systemic (Qs) flow ratio greater than 1.5:1. The Madrid report compared 192 adults after surgical ASD closure, with 94 non-operated adults with an ASD. In the first two studies, patients with sustained (symptomatic) or chronic (>3 months) atrial flutter/fibrillation based on a 12-lead ECG and/or 24-hour Holter monitoring were included. The

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mean age at surgical closure of the ASD was 42 (range 18–79) years. In the Toronto study, 19% of patients had documented atrial flutter/fibrillation pre-operatively [4], while in the Berlin study this incidence was 27% [7]. The mean age of patients with ASD and atrial flutter/fibrillation was 59 ± 11 years in Toronto, and was 57 ± 10 in Berlin. Both reports found an increase in atrial flutter/fibrillation with age. Atrial flutter/fibrillation was rarely seen in patients younger than 40 years. In the Berlin study 3/55 (5.4%) of the patients who had atrial flutter/fibrillation, presented with a stroke before the ASD was closed. In two of those three patients the stroke led to further cardiac investigations and ultimately to the diagnosis of an ASD [7]. Only one patient with an ASD out of the whole group (0.5%) had a stroke while being in sinus rhythm.

The incidence of arrhythmia post-operatively was not related to the technique of surgical repair, ie a direct suture versus patch closure of the ASD [4, 7]. In the Toronto study, atrial flutter and AF were considered together [4]. At a follow-up longer than 3 years after defect closure, 60% of the patients continued to have flutter/fibrillation. Due to ongoing arrhythmia and an enlarged left atrium, the risk of embolism and stroke still persisted, indicating long-term prophylactic anticoagulation, recommended to prevent cerebro-vascular accidents. Only 40% of patients in whom an ASD was closed successfully reverted to sinus rhythm. In addition, new onset flutter/fibrillation developed in another 5/67 (7.4%) patients older than 40 years. Despite documentation of successful ASD closure and adequate anticoagulation, in all but one of those five patients this led to a stroke post-operatively. The Toronto group concluded from their data, that mere closure of the defect may not be enough to prevent strokes, and that an additional Cox-Maze operation could be considered in the future [4].

In the Berlin study, the patients with atrial arrhythmia were divided into 2 groups, one with atrial flutter and another with AF [7]. Significant differences with regard to the prevalence and prognosis were found between the 2 arrhythmias. In patients younger than 40 years of age, none had AF pre-operatively. While a transient short lasting-period of atrial fibrillation was present between the 3rd and 5th post-operative day in 2 patients (2%), no patient retained AF in the long-

term follow-up. In middle-aged patients (40–60 years old) the pre-operative prevalence of atrial flutter and fibrillation was similar. In patients older than 60 years, AF was the predominant arrhythmia [7] (Table 1). It is important to consider that the prognosis of atrial flutter compared to AF is entirely different. Atrial flutter converted to sinus rhythm late after surgical repair in 10/18 patients (55%), whereas only 4/25 (12%) patients with pre-operative AF converted to sinus rhythm ($p < 0.04$) after ASD closure [7]. As AF is the predominant tachy-arrhythmia in older patients, it is not surprising that older patients have a worse prognosis than younger patients with respect to reverting to sinus rhythm after surgical ASD closure. One may conclude from the Toronto and Berlin studies that in patients with AF, closure of the ASD alone may not be enough to prevent future atrial rhythm disturbances. If arrhythmia persists or appears after ASD closure, we recommend continuous prophylactic anticoagulation. In an attempt to influence and reduce ongoing fibrillation and the need for continuous prophylactic anticoagulation to prevent cerebro-vascular accidents, anti-arrhythmic procedures should be considered at the time of surgical defect closure.

These results are strengthened by the Madrid study, in which 192 adults having already undergone ASD closure were compared to 94 patients with a non-operated ASD [8]. The incidence of AF was similar in surgical and non-surgical patients, and was related to advanced age, left atrial size, and the degree of mitral and tricuspid regurgitation. Only age >25 years at the time of ASD closure was a predictor of AF in the surgical group [8]. The Madrid group concluded that closure of an ASD before adulthood should be encouraged, thereby reducing the incidence of post-operative AF, and consequently eliminating the need for a concomitant anti-arrhythmic procedure. They did not comment on the need for an anti-arrhythmic procedure concomitant to ASD closure when patients present later than 25 years of age.

The 3 studies do not make a distinction between paroxysmal flutter/fibrillation, chronic atrial arrhythmia, and symptomatic atrial flutter/fibrillation, with respect to an eventual need for ASD closure and reduction in arrhythmia prevalence, or a concomitant anti-arrhythmia procedure. Whether or not this distinction is essential is unknown and unanswered by the literature.

Table 1

Incidence of arrhythmia before and after atrial septal defect closure and its regression in the Berlin study group [7].

Arrhythmia	18–40 years of age (n = 101)	40–60 years of age (n = 83)	>60 years of age (n = 27)
Atrial flutter before surgery	1 (1%)	12 (15%)	5 (19%)
Atrial fibrillation before surgery	0	12 (15%)	16 (61%)
Atrial flutter after surgery	0	7 (9%)	3 (11%)
Atrial fibrillation after surgery	0	8 (10%)	13 (48%)
Reverted to sinus rhythm from pre-operative atrial flutter	1 (100%)	5/12 (42%)	2/5 (40%)
Reverted to sinus rhythm from pre-operative atrial fibrillation	Na.	4/12 (33%)	3/16 (19%)

Na= Not applicable

Concomitant anti-arrhythmia surgery

Although it seems reasonable to assume that some form of concomitant anti-arrhythmic surgery will be needed in patients with chronic atrial dysrhythmias undergoing surgical ASD closure [9–15], it is still debatable which type of anti-arrhythmic procedure is warranted. In addition, it is unclear whether this is required prophylactically in older patients without chronic arrhythmia at the time of surgery. Large studies have shown that for AF in adults, with or without the association of congenital heart disease, a left-sided Cox-Maze III is the appropriate procedure with the lowest recurrence [12–14]. Various smaller studies also report success with the less extensive right-sided Maze procedure to treat AF, although their follow-up is much shorter [9–11, 15]. The one group that performed both right-sided and/or left-sided Cox-Maze III procedures doubted that a right-sided Maze procedure alone would be sufficient to prevent or to treat AF [14]. Mavroudis et al. have demonstrated the efficiency of performing the limited right-sided Maze in patients with atrial flutter or atrial re-entry tachycardia, but recommend adding a Cox-Maze III when AF is present [13].

Currently, there is no consensus regarding which anti-arrhythmic procedure should be per-

formed in patients with various types of established atrial arrhythmia. Even in the absence of an intra-cardiac defect such as an ASD there is no general procedure, although most authors agree that some sort of surgical anti-arrhythmia surgery is indicated [9–15]. In older patients without arrhythmia at the time of ASD closure, but at risk of developing post-operative atrial rhythm disturbances, the dilemma to perform a prophylactic anti-arrhythmia procedure remains. Prospective randomised trials are required to answer these delicate questions, since an additional right-sided Maze or a left-sided Cox-Maze III procedure are not without additional surgical risk. A right-sided procedure does add to cardiac ischemic time, as it can be performed without aortic cross-clamping, and may be enough for atrial flutter or atrial re-entry tachycardia [13]. The left-sided Cox-Maze III is a more demanding procedure, and involves additional ischemic time for the heart, which is not without potential negative consequences for cardiac recovery. Although the left-sided Cox-Maze III has been performed without additional post-operative morbidity or mortality and is the most reliable surgical procedure to treat established AF [12, 13], the advantages and disadvantages must be weighed before recommending it routinely.

Comment

Data from the literature and our own study demonstrated that 1) the incidence of pre-operative atrial flutter/fibrillation increases with age, 2) that atrial flutter is the predominant arrhythmia in younger patients, and AF is more common in older patients, and 3) that the probability of atrial flutter to return to sinus rhythm after removing right heart volume overload by closure of the defect is higher than that of AF. These data support the notion that patients with AF should be managed differently than patients with atrial flutter. As the results of ablation procedures in fibrillation are poorer than those of flutter ablation, an operative left-sided Cox-Maze III procedure concomitant to ASD closure should be considered in selected patients with AF [12, 13].

The clinical importance of atrial flutter/fibrillation mainly lies in its association with stroke. In certain studies, up to 22% of late deaths after surgical ASD closure were caused by stroke, and none of these patients had sinus rhythm [16–18]. The mechanisms leading to atrial flutter in ASD include atrial dilatation, increased atrial pressure,

and conduction disturbances [19, 20]. If the underlying problem, ie the atrial shunt is effectively removed, dilatation may regress and the incidence of flutter may decrease [19]. It is obvious that the probability for the atrial size to return to normal is smaller, the longer the defect remains untreated [21]. Thus the best prevention of atrial flutter in patients with an ASD is earlier diagnosis and treatment of the defect.

As the etiology of AF is less well understood than that of atrial flutter, the diagnostic approach of the rhythm disturbance and the hemodynamic abnormality is more complicated. The Framingham study demonstrated that the incidence of new onset of AF, which was 0.5% in the 5th decade of life, doubled with each decade of age, independently of the increasing prevalence of known predisposing conditions [22]. Thus, in some patients, atrial fibrillation may not necessarily be caused by an ASD. This has to be taken into account before new therapeutic strategies such as a Cox-Maze III procedure can be recommended, concomitant to closure of the ASD.

Conclusions

Adult patients with an ASD and atrial tachyarrhythmias benefit from defect closure. Closure of the defect may lead to regression of atrial flutter, whereas in older patients (>25 years) with AF, restoration of normal hemodynamics alone may not be sufficient [8]. Most surgical groups are inclined to perform a right-sided Maze concomitant to ASD closure in patients with atrial flutter [9–11, 13–15]. Others recommend a left-sided Cox-Maze III for patients with AF [12, 13], although this has yet to be validated with randomised studies. The

role of prophylactic anti-arrhythmic procedures in older patients with an ASD is currently unknown.

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