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Reinterventions in patients with congenital aortic stenosis and a commissurotomy

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ABSTRACT

Background In congenital aortic stenosis (CAS), commissurotomy is an option in patients not suitable to receive a valve prosthesis. However, there is often a need for future additional interventions on the aortic valve. The fate of the aortic valve is, however, essentially unknown. This study reports the need for reinterventions after surgical commissurotomy, based on a national register.

Materials and methods The national register on congenital heart diseases (CHDs) was searched for patients with CAS, simple or associated with other CHD and an index commissurotomy with later data from follow-up.

Results 300 patients with CAS and an index commissurotomy (mean age at the operation 7.4±7.8 years, 72.4% males) were identified. After an observation time of 27.4±10.0 years, 54.7% of the patients had a reintervention that occurred 14.2±10.1 years after the index operation. The cumulative incidence of reintervention was approximately 25% 10 years after and 60% 30 years after the index intervention. The prevalence of left ventricular hypertrophy (LVH) was higher among those that needed reintervention (41.3 vs 26.8%, p=0.023). Furthermore, eight patients died with a cumulative incidence of 7% 30 years after the index intervention, where most were (7/8) without reintervention (p=0.025). There were no additional important differences between patients with and without reintervention. The prevalence of left ventricular dysfunction and New York Heart Association class >1 was low.

Conclusion Most (54.7%) patients with a commissurotomy, more than half of them within 30 years and eventually all will need a reintervention. This is important information to patients, especially for women in childbearing age. The higher prevalence of LVH in the group with reintervention needs attention during follow-up. Furthermore, those without reintervention, for unknown reasons, had a higher mortality. Our data strengthen the arguments for close outpatient follow-up among patients with a previous commissurotomy.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Congenital aortic stenosis is a life-threatening condition in children, often leading to heart failure and death. Commissurotomy is an option for patients who are not suitable for valve prosthesis due to anatomical issues or young age. However, there are only a few smaller studies with long-term outcomes, and this is one of the largest studies with a longer follow-up period to assess outcomes.

WHAT THIS STUDY ADDS

⇒ Over time, almost all patients treated with commissurotomy will require at least one additional aortic valve intervention. Commissurotomy for congenital aortic stenosis is not a definitive treatment but rather a bridge to a final intervention. The study also provides a detailed overview of relevant surgical and catheter-based interventions performed on patients with congenital stenosis of the aorta valve over a period of 50 years.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ Patients and their parents need to be informed that commissurotomy is only a bridge to further interventions and not a final treatment, even if reoperation may take decades. The study also underscores the importance of lifelong regular follow-up to identify valve deterioration and the need for reoperation.

INTRODUCTION

In general, aortic stenosis stands out as the most prevalent valve disease and with congenital aortic stenosis comprising roughly 3–6% of congenital heart defects.[1 2] The severity spectrum varies; neonates and young infants may present with congestive heart failure, whereas older children and adolescents most often are asymptomatic.[3] Intervention is necessary for symptomatic patients and for most with severe left ventricular outflow tract obstruction.[3] If the patient is too small for a prosthetic valve, transcatheter balloon valvotomy, open commissurotomy or Ross procedure are therapeutic alternatives.[4 5] In the Ross procedure, the pulmonary valves are transposed to the aortic position and a

Table 1 Patient characteristics divided into total cohort, those with no reintervention and those without reintervention.

	All (300)	No reintervention (n=136)	Reintervention on the aortic valve (n=164)	P value
Sex, male, n (%)	217 (72.3)	97 (71.3)	120 (73.2)	0.72
Age at primary commissurotomy, year	7.4±7.8 (n=300)	8.2±8.3 (n=136)	6.7±7.3 (n=164)	0.11
Age at first new intervention, year		N.A.	20.9±13.2 (n=164)	N.A.
Age at data study end, year	34.9±11.3 (n=300)	31.9±9.8 (n=136)	37.3±11.9 (n=164)	**<0.001**
Age at death, year	33.2±12.9 (n=8)	33.6±13.8 (n=7)	30.8 (n=1)	0.86
Height, cm	174.2±10.9 (n=272)	173.8±11.3 (n=124)	174.5±10.6 (n=148)	0.60
Weight, kg	77.0±17.0 (n=242)	76.6±17.4 (n=111)	77.3±16.7 (n=131)	0.76
Body mass index, kg/m^2	25.2±4.9 (n=241)	25.1±4.7 (n=110)	25.3±5.1 (n=131)	0.78
Systolic blood pressure, mm Hg	122±16 (n=277)	121±15 (n=122)	124±16 (n=155)	0.14
Diastolic blood pressure, mm Hg	73±10 (n=277)	72±10 (n=122)	73±10 (n=155)	0.27
EuroQol Group 5D visual analogue scale	82.7±16.4 (n=179)	84.3±14.7 (n=79)	81.4±17.7 (n=100)	0.24
Smoking: non-smoker/previous smoker/smoker, n (%)	88.2/5, 5/6, 2% (n=289)	88, 5/4, 6/6, 9 (n=131)	88, 0/6, 3/5, 7 (n=158)	0.91
Symptoms, yes, n (%)	40 (13,4) (n=298)	18 (13,3) (n=135)	22 (13,5) (n=163)	0.97
Pacemaker/implantable cardioverter defibrillator, yes, n (%)	14 (4.7) (n=298)	3 (2.2) (n=135)	11 (6.7) (n=163)	0.055
Anticoagulant drugs, yes, n	85 (28.4) (n=299)	3 (2.2) (n=133)	82 (50.0) (n=163)	**<0.001**
Arterial hypertension, yes, n (%)	69 (34) (n=203)	27 (32.1) (n=84)	42 (35.3) (n=119)	0.64
Left ventricular hypertrophy, yes (%)	78 (35.0) (n=223)	26 (26.8) (n=97)	52 (41.3) (n=126)	**0.02**
Normal left ventricular function (ejection fraction >50%), yes, n (%)	228 (95.4) (n=239)	97 (96.0) (n=101)	131 (94.9) (n=138)	0.69
New York Heart Association class I, n (%)	214 (88.8) (n=241)	90 (90.9) (n=99)	124 (87.3) (n=142)	0.39

For each variable, the number of observations is presented. Bold p-values indicate p<0.05.

pulmonary homograft is placed in the pulmonary position. Additionally, approximately 15–20% of individuals with congenital aortic stenosis also present with associated heart defects, such as coarctation of the aorta and dilatation of ascending aorta, among other less common defects.[6]

The size of the aortic valve annulus increases with age, making commissurotomy more suitable for neonates and young infants.[4] Conversely, the Ross procedure is typically reserved for older children and adolescents, who are still too small to receive a mechanical valve prosthesis.[7]

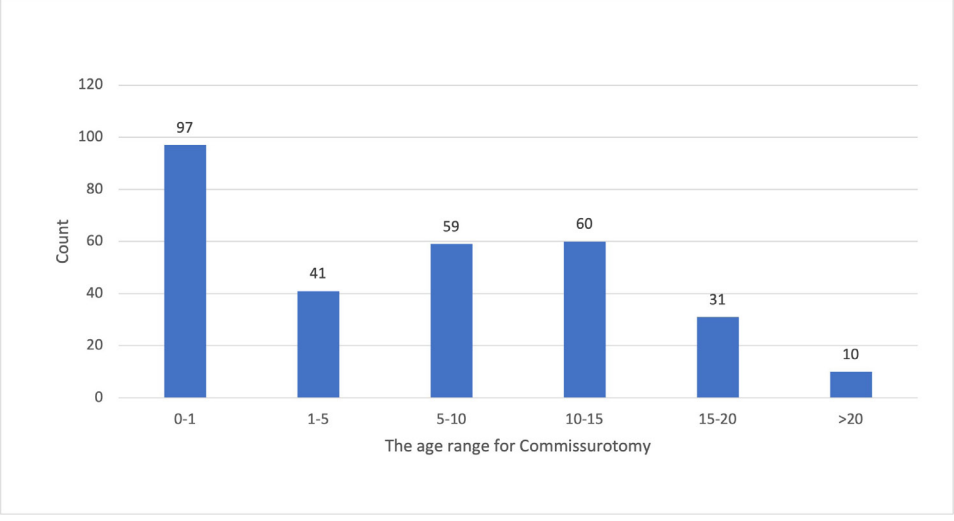

Figure 1 Age in years at commissurotomy. The total number of patients is 300. The mean age at the index intervention was 7.4±7.8 years.

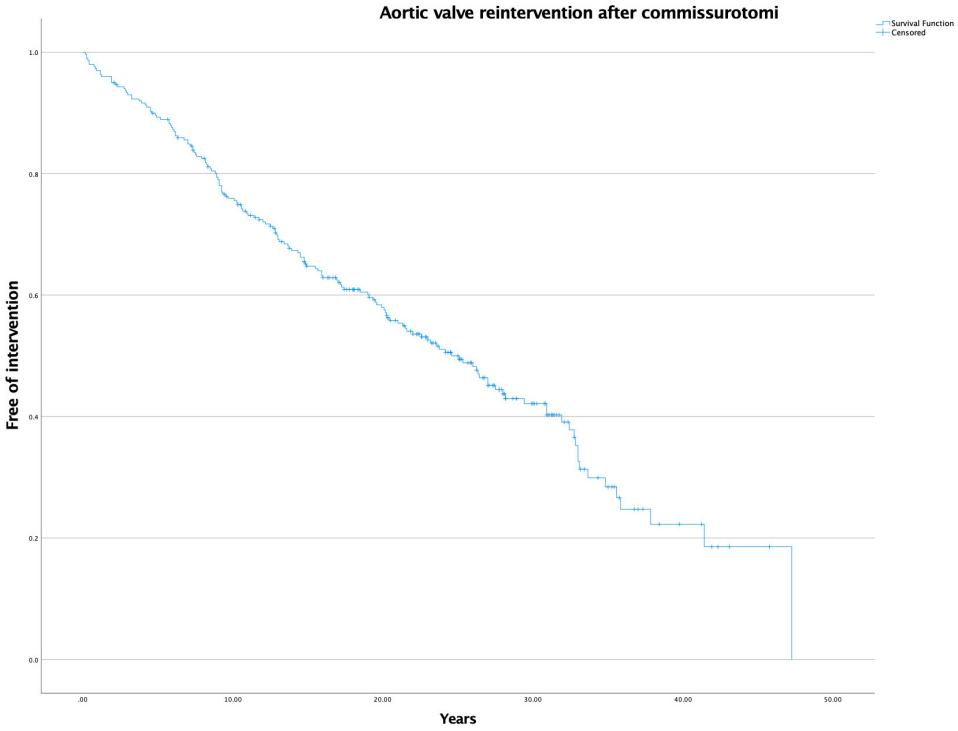

Figure 2 Kaplan-Meier curve showing aortic valve reinterventions following commissurotomy, with a mean observation time of 27.4±10.0 years from the index operation to study end. The reintervention occurred 14.2±10.1 years after the index operation.

After open commissurotomy, reinterventions appear common, but their prevalence is not well known as well as the total burden of other interventions in this population.[8 9] Previous research on commissurotomy typically involves a limited number of patients and targets specific age groups.[10 11] In this population-based register study, the entire Swedish population is considered in all age groups above 18 years of age. The aim of this investigation was to study the need for reinterventions and prognosis in adults with a previous commissurotomy on the aortic valve.

MATERIALS AND METHODS

The SWEDCON registry

The national register of congenital heart disease (CHD) contains four parts: (1) fetal echocardiography, (2) congenital heart surgery, (3) paediatric cardiology (patients <18 years and all heart diseases) and (4) adult (<18 years) congenital heart disease (ACHD, ie, only structural CHD excluding, eg, cardiomyopathies and hereditary arrhythmias). In this study, only the ACHD part of the register was used. This part started in 1992 and since 1998 covers almost the whole population of Sweden with CHD and over 18 years of age.[12] Data entry is done by the seven clinics specialised in ACHD and clinics in 17 county hospitals. In the register, there is information about the diagnosis, interventions, demographics and medication. Information about the cardiovascular symptoms, New York Heart Association (NYHA) class and echocardiographic data, including left ventricular function, left ventricular hypertrophy (LVH) (defined as any hypertrophy at echocardiography), blood pressure, quality of life and physical activity level.

Patient selection

When data were extracted on 6 October 2017, the register contained 300 patients older than 18 years of age and who had CAS and undergone commissurotomy at any age and at least one clinical visit registered. The first commissurotomy in the register was performed in 1964 and the last one in 2015. CAS was defined as congenital aortic valve stenosis, with or without associated CHD, that needed intervention, and that the primary intervention was commissurotomy.

Statistical analysis

The statistical analysis was performed using IBM SPSS Statistics, V.28.0 (IBM, Armonk, New York, USA). Data are presented as means±SD. Frequencies are presented as numbers with percentages. For comparisons, Student's t-test was used for continuous variables and χ^2 test was used for categorical variables. The probability of survival was assessed using Kaplan-Meier curves with log-rank tests. The null hypothesis was rejected on p values <0.05.

RESULTS

In total, 300 patients with an index commissurotomy (mean age at surgery 7.4±7.8 years, 72.3% males) were identified (mean age at data extraction 34.9±11.3 years) (table 1).

The age distribution at the index operation is shown in figure 1. After a mean observation time of 27.4±10.0 years

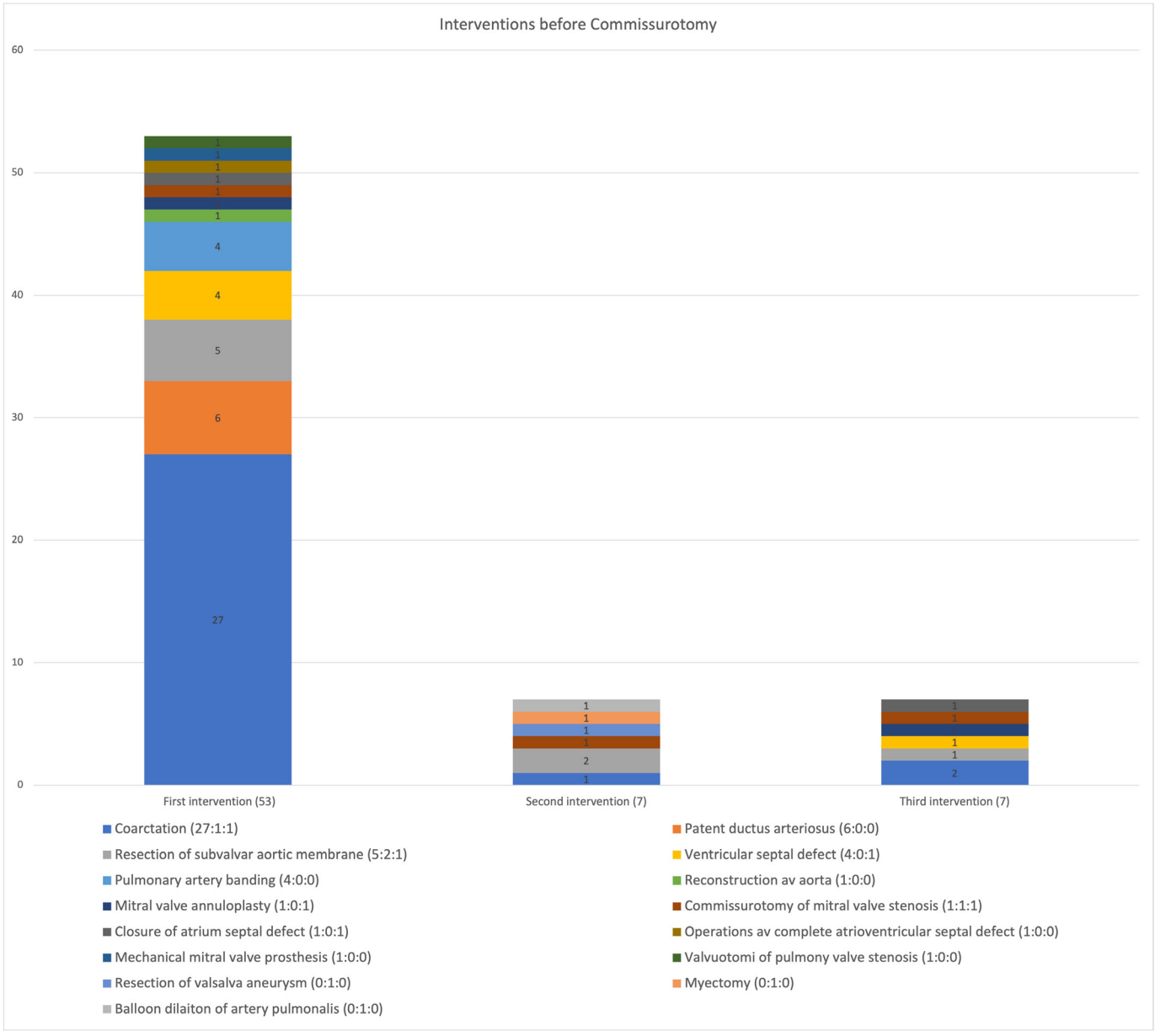

Figure 3 The number of interventions prior to the index operation. The majority required one intervention, while others needed two or three reinterventions. The first number represents the count for the first intervention, etc. The most common heart defect associated with congenital aortic stenosis in our data is coarctation.

from the index operation to study end, 164 (54.7%) of the patients had a reintervention on the aortic valve. The reintervention occurred 14.2±10.1 years after the index operation. Patients with a reintervention were older at data extraction (37.3±11.9 vs 31.9±9.8, p<0.001) and were more often treated with anticoagulants at the last clinic visit (table 1).

The prevalence of LVH was higher among those that needed reintervention (41.3 vs 26.8%, p=0.023). Eight patients died (five men and three women) during observation in the register with a cumulative incidence of 7% at 30 years after the index intervention, most (7/8) without reintervention (p=0.025). One patient needed heart transplantation after several reoperations. There were no important demographic or clinical differences between patients with and without reintervention. The prevalence of left ventricular dysfunction and NYHA class >1 was low in both groups (table 1).

The cumulative incidence of first reintervention on the aortic valve was approximately 25% at 10 years after and 60% 30 years after the index intervention (figure 2).

There were 164 first reinterventions on the aortic valve, 64 due to aortic regurgitation, 39 aortic stenosis, 1 infectious endocarditis and 60 unknowns. They were treated with aortic valve replacement with mechanical prosthesis (n=40) or bioprosthesis (n=30), a composite aortic valve/aortic graft (n=25), Ross operation (n=30), recommissurotomy (n=34), valvuloplasty due to aortic regurgitation (n=1), balloon dilation (n=3) and unknown (n=1). There were 44 second interventions and 5 third interventions

Table 2 Number of interventions performed excluding aortic valve surgery.

Type of interventions	Number of interventions (n=51)
Aortic reconstruction	9
Subaortic membrane resection	9
Mitral valve annuloplasty	7
Pulmonary homograft	6
Percutaneous pulmonary artery stent	4
Closure of ventricular septal defect	3
Mechanical mitral valve replacement	<3
Atrial fibrillation surgery	<3
Mechanical pulmonary valve replacement	<3
Bioprosthetic pulmonary valve replacement	<3
Pulmonary balloon dilation	<3
Bioprosthetic mitral valve replacement	<3
Mitral valve commissurotomy	<3
Closure of atrial septal defect	<3
Heart transplantation	<3
Coronary artery bypass graft	<3
Unknown	<3

To ensure patient anonymity, interventions fewer than three are indicated as <3.

on the aortic valve (figure 3). In addition, there were 51 interventions that did not involve the aortic valve (table 2).

Prior to commissurotomy, there were 53 first interventions, 7 second interventions and 7 third interventions (figure 4).

DISCUSSION

Here we show that new operations eventually are needed in most patients with a previous commissurotomy due to congenital aortic stenosis. On a group level, many procedures are needed both before and after commissurotomy, often caused by other concomitant CHD. The long-term outcome is good both in terms of ventricular function and NYHA class. Possible problems are LVH, more commonly seen in those with a reintervention on the aortic valve. Mortality is rare, but for unclear reasons more common in those without reintervention. Thus, reoperations are prevalent both on the aortic valve and for other reasons. Therefore, patients should be periodically monitored and be informed that a reintervention on the aortic valve is likely in the future, unfortunately in an uncertain perspective of time.

More than half of the patients with a previous commissurotomy needed reinterventions on the aortic valve. According to estimations in the Kaplan-Meier analysis, most of them will eventually need aortic valve surgery. Thus, commissurotomy should be considered a palliative intervention as most patients will need future interventions. Furthermore, many patients have concomitant CHD and need interventions for other lesions not involving the aortic valve. Indeed, more than 60% of patients needed three or more interventions (Figure 5). This shows that patients with a previous commissurotomy are very likely to have a high number of thoracotomies, in a few cases close to the limit of what is possible. There are few previous studies on the follow-up after commissurotomy on the aortic valve. In children followed to the age of 14 years, approximately half of them needed a reintervention on the aortic valve within 10 years.[10] This is even higher than the cumulative incidence of 30% over the same period of time presented by us. It is unclear why the results differ, but it may be that prognosis in the term reinterventions is better in older patients. Nevertheless, as our study has a long observation time and contains older patients, most patients with a previous commissurotomy appear to need a new intervention on the aortic valve.

The number of deaths was low and mainly occurred in patients without reinterventions. The number, however, is low and may differ between groups just by chance. Still, it cannot be excluded that some patients were severely affected by the heart disease and therefore not suitable for surgery. Another finding was LVH among those with reinterventions. This can be due to more severe valvular disease prior to interventions, which is a concern and should be considered during follow-up.

A fourth of the patients with reinterventions on aortic valve received a mechanical prosthesis on the first reintervention. This is also reflected by the higher prescription of anticoagulant drugs for patients with reinterventions. In this context, it must be remembered that mechanical valves and anticoagulation are associated with long-term complications such as embolic stroke and bleeding.[13] It was, however, beyond the scope of our study to analyse that type of long-term complications. Nevertheless, in young patients, both children and adults, with aortic valve disease, a valve prosthesis without the need of anticoagulation may be considered.[14–16] In fact, 50% of the patients in our material received a non-mechanical alternative at their first reintervention.

This study has some limitations. First, it is a register study and thus limited to the patients included in the register. Second, there are potentially patients with commissurotomy who died before the age of 18 years and thus not included in the register, that is, immortal time bias. Third, there are no detailed data on the valve morphology, limiting the possibility to draw conclusions based on valve morphology and function. Fourth, although we are restricted to the data in the register, our data are national, multicentre and, in the context of CHD, contain a substantial number of observations, also shown in several of our studies in adults with CHD.[17–19] This may compensate for the potential limitations listed above.

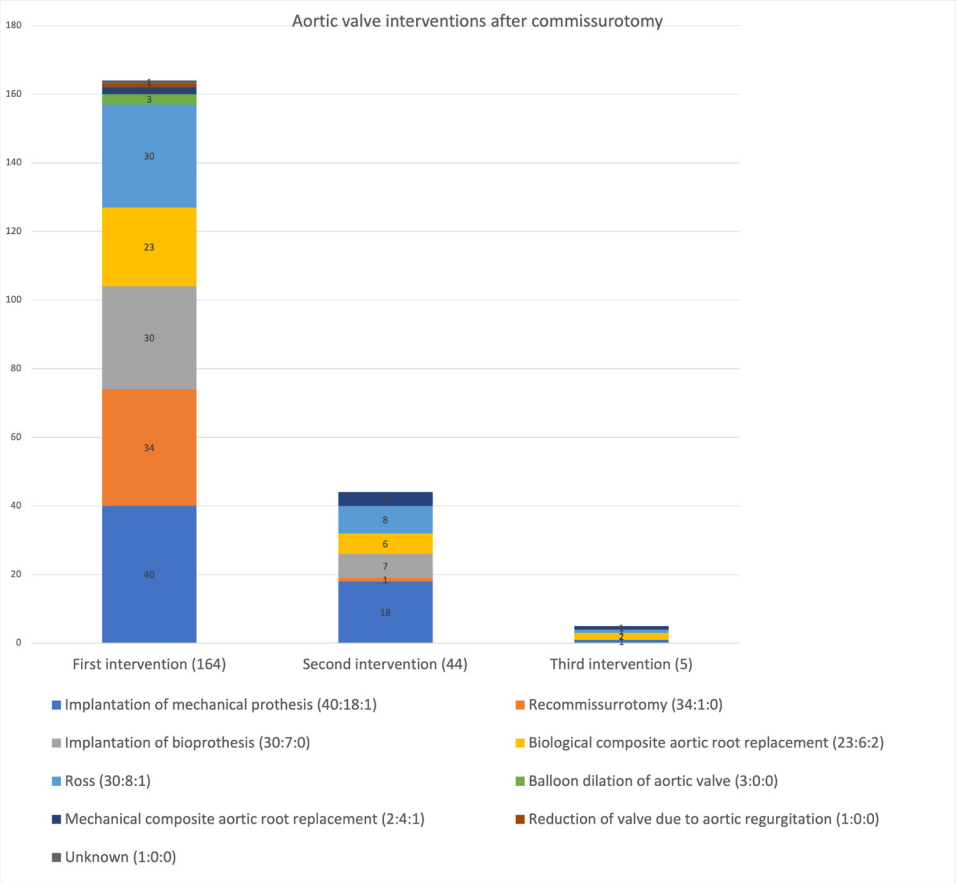

Figure 4 Aortic valve reinterventions after commissurotomy: 164 patients underwent at least one reintervention, 44 underwent at least two reinterventions and 5 underwent at least three reinterventions. The numbers following the interventions indicate the quantity of interventions. The first number represents the count for the first intervention, etc. The most common intervention after commissurotomy is the implantation of a mechanical aortic valve. Recommissurotomy is also common.

CONCLUSION

Most patients with a commissurotomy in childhood, eventually almost 100%, will need a reintervention, that is, important information to patients, especially for women in childbearing age. The higher prevalence of LVH in the group with reintervention needs attention during

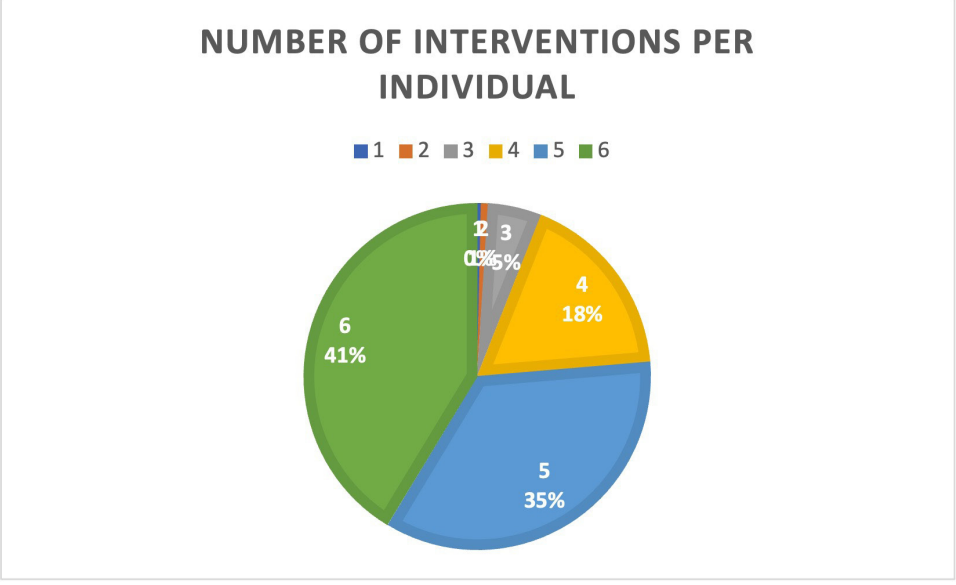

Figure 5 Number of interventions per individual. At least 71% underwent more than one intervention. Two individuals in our data underwent six interventions each, consisting of five thoracotomies and one endovascular treatment.

follow-up. Furthermore, those without reintervention, for unknown reasons, had a higher mortality. Our data strengthen the arguments for close outpatient follow-up among patients with a previous commissurotomy.

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Competing interests None declared.

Patient consent for publication Not applicable.

Ethics approval This study involves human participants and was approved by the Regional Ethics Review Board in Umeå, Sweden, 08-218M and 2012-445-32M. This is a register study with approved ethics review, so no informed consent was required.

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Data availability statement Data may be obtained from a third party and are not publicly available.

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