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# Outcomes following aortic valve procedures in 201 complex congenital heart disease cases - Results from the UK National Audit

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Key question: What are the outcomes following aortic valve procedures in complex congenital heart disease?

Key findings: Aortic valve procedures in complex congenital cases can be performed with good outcomes outside neonatal ages.

Take-home message: In complex congenital heart disease aortic valve procedures carry a risk, but mortality is still higher than in other scenarios.

Central figure legend: Registry outcome data in complex aortic valve procedures can inform clinical decision and counselling.

**Abstract and key words**

**Objective:** Some patients with complex congenital heart disease (cCHD) also require aortic valve (AoV) procedures. These cases are considered high risk but their outcome has not been well characterised. We aim to describe these scenarios in the current practice, and provide outcome data for counselling and decision making. .

**Methods:** This was a retrospective study using the UK National Congenital Heart Disease Audit data on cCHD patients undergoing aortic valve replacement (AVR), balloon dilation (BAV) or surgical repair (SAV) between 2000-2012. Coarsened exact matching was used to pair cCHD with patients undergoing AoV procedures for isolated valve disease.

**Results**: A total of 201 patients with a varied spectrum of cCHD undergoing 242 procedures were included, median age 9.4 years (1 day-65 years). Procedure types were: BAV (n=31, 13%), SAV (n=57, 24%) and AVR (n=154, 63%). Mortality at 30 days was higher in neonates (21.8% vs 5.3%, p=0.02).

Survival at 10 years was 83.1%, freedom from AVR 83.8% and freedom from BAV/SAV 86.3%. Neonatal age (p<0.001), single ventricle (p=0.08), concomitant Fontan/Glenn (p=0.002) or aortic arch procedures (0.02) were associated with higher mortality. cCHD patients had lower survival at 30 days (93% vs 100%, p=0.003) and at 10 years (86.4% vs 96.1%, p=0.005) compared to matched isolated AoV disease patients.

**Conclusions:** AoV procedures in cCHD can be performed with good results outside infancy, but with higher mortality than in isolated AoV disease. Neonates and patients with single ventricle defects, especially those undergoing concomitant Fontan/Glenn, have worse outcomes.

**Key words:** aortic valve replacement; balloon aortic valvotomy; congenital heart disease; tetralogy of Fallot; transposition of great arteries; single ventricle.

Glossary of abbreviations

AoV – aortic valve

AVR – aortic valve replacement

AVSD – atrioventricular septal defect

BAV – balloon aortic valvoplasty

cCHD – complex congenital heart disease

CCTGA – congenitally corrected transposition of great arteries

DORV – double outlet right ventricle

HLHS – hypoplastic left heart syndrome

IAA – interrupted aortic arch

NCHDA – National Congenital Heart Disease Audit

NICOR – National Institute for Cardiovascular Outcomes Research

PA-IVS – pulmonary atresia with intact septum

PA-VSD – pulmonary atresia with ventricular septal defect

SAV – surgical aortic valve repair

SV – other single ventricle physiology

TGA – transposition of the great arteries

TGA-IVS – transposition of great arteries with intact septum

TGA-VSD – transposition of great arteries with ventricular septal defect

ToF – tetralogy of Fallot

## INTRODUCTION

Congenital aortic valve (AoV) lesions, the most common congenital heart disease (CHD), can be associated with complex CHD (cCHD), and require multiple interventions throughout the patients' lives, involving several cardiac structures.[1] Outcomes of AoV interventions are well described in the general population, but data are limited in patients with concomitant cCHD.[2] Also unknown is how they compare to those with isolated AoV lesions, with no specific mention in the guidelines on the subject.[3, 4] This can lead to a lack of evidence-based counselling and decision-making.

Previous studies looked at balloon aortic valvuloplasty (BAV), surgical aortic valve repair (SAV) and aortic valve replacement (AVR), but exclude or have few cases with cCHD.[2, 5–8] Even when the association with AoV disease is well documented, such as hypoplastic left heart syndrome (HLHS),[9, 10] transposition of the great arteries (TGA)[11–14] or interrupted aortic arch (IAA)[15, 16] the details on AoV intervention are limited. Data are even more scarce in right heart or single ventricle defects, where only case reports or small series are available.[17–20]

A multicentre registry, such as the United Kingdom National Congenital Heart Disease Audit (NCHDA) is well equipped to show the picture of AoV intervention in cCHD.

This study aims to: 1. describe the population of cCHD patients undergoing AoV procedures and their outcomes after BAV, SAV and AVR; 2. identify predictors of mortality and reintervention; and 3. compare outcomes in the cCHD and isolated AoV disease, in order to offer outcome data to be used as future reference in counselling and decision making on the topic of AoV procedures in complex CHD.

## PATIENTS AND METHODS

### *Ethical statement*

The NICOR holds approval to use patient data for research purposes. The data request for this manuscript was processed and approved by the NICOR Research Board. The need for patient-level consent to participate in this retrospective study with anonymized data was waived by the NICOR Research Board.

***The dataset***

The National Institute for Cardiovascular Outcomes Research (NICOR) collects validated data on cardiac procedures from all UK units in the NCHDA, described previously.[6] Linkage with survival registries of Northern Ireland and Scotland cannot be performed, while some patients are from overseas. This resulted in 12.4% (n=25) of patients not having follow-up data beyond 30 days, being censored at the time of discharge. Procedure indication and timing were established by the multidisciplinary teams at each centre. Children are defined as below 16 years of age.

***Patient selection and classification***

All available data on CHD patients undergoing BAV, SAV or AVR between April 2000 and March 2012 were selected (n=4328). cCHD included the following, adapted from Warnes et al. and Hoffman et al:[1, 21] absent pulmonary valve, aortic valve hypoplasia, atrioventricular septal defect (AVSD), congenitally corrected transposition of great arteries (CCTGA), double outlet right ventricle (DORV), Ebstein anomaly, hypoplastic left heart syndrome (HLHS), interrupted aortic arch (IAA), other single ventricle physiology (SV), pulmonary atresia with intact septum (PA-IVS), pulmonary atresia with ventricular septal defect (PA-VSD), tetralogy of Fallot (ToF), transposition of great arteries with intact septum (TGA-IVS), transposition of great arteries with ventricular septal defect (TGA-VSD) and totally anomalous pulmonary venous connection. Patients with common arterial trunk (n=52) were not included, as the truncal valve was considered to be anatomically different, and warrants separate study. Finally, transcatheter AVR, less than 10 cases, was excluded.

This resulted in a total of 201 patients with cCHD undergoing 241 AoV procedures, detailed in Table 1. The first AoV procedure in the database for each patient was the index procedure. Reinterventions were defined as AoV procedures, surgical or transcatheter, following the index procedure and divided into repair (BAV/SAV) or replacement (AVR) subgroups. Early events were defined as occurring within 30 days from the index procedure.

***Statistical Analysis***

Frequencies are given as numbers and percentages, continuous values as median (inter-quartile range). Population characteristics were compared using the Mann-Whitney U (continuous 2 groups), Kruskall-Wallis (continuous >2 groups), and Fisher's exact test (proportions). Survival and freedom from reintervention are estimated using the Kaplan-Meier method, considering the time from the index procedure up to the first event of each type (death, AVR, BAV/SAV) and presented as percentage and 95% confidence interval (CI).

To identify predictors for mortality and reintervention we used univariable Cox regression (more details in Supplementary Material). To investigate the impact of cCHD on outcomes, we compared this group to a matched population with isolated AoV. We selected patients undergoing AoV interventions for purely isolated congenital AoV, from the same database (n=1665). We then used a coarsened exact matching algorithm, with 1-to-1 pairing and random pruning of pairs to matched the two groups.[22] Variables used in the matching were: age (continuous), age group (categorical), gender, aortic valve disease type (stenosis, regurgitation, unknown) and procedure year (continuous). This resulted in 133 cCHD-isolated AoV disease pairs (multivariate L1 distance pre-matching=0.84, post-matching=0.18, lower means less imbalance). Statistical analyses were done using STATA/SE 12 (StataCorp LP, College Station, TX).

## RESULTS

A total of 201 patients with cCHD (1 day-65 years, median 11.4 years) underwent 241 AoV procedures, 18% BAV (n=43), 26% SAV (n=62) and 56% AVR (n=136). Of the 136 AVR procedures, 46% (n=63) were mechanical AVR, 11% (n=15) Ross AVR, 6% (n=8) Ross-Konno AVR, 4% (n=6) homograft AVR and 2% (n=3) bioprosthesis AVR, while 30% (n=41) were unspecified AVR; 20% (n=26) had a concomitant aortic root/ascending aorta replacement. Demographic, clinical and procedural data are detailed in Table 1.

The diagnoses by procedure type (AVR/BAV/SAV) are listed in Table 2. Left-sided associated defects were more common in neonates and infants, right-sided associated defects were more frequent in adults (Figure 1). AoV disease type was documented in 78% of patients (n=156); stenosis was more common in left heart cCHD, while regurgitation was more common in right heart or transposition type cCHD (Figure 2).

From the 241 AoV interventions, 97 (40%) had concomitant surgical (n=94) or transcatheter (n=3) procedures performed (Supplementary Table 1). The most common concomitant surgical procedures were: pulmonary valve/conduit replacement (n=30), VSD closure (n=19), pulmonary arterioplasty (n=14), aortic arch surgery (n=11) and Fontan/Glenn (n=11).

### *Early outcomes*

Early (30 day) mortality and reintervention in n=213 AoV procedures with follow-up beyond 30 days were 7% and 1.8%, respectively. Early mortality was higher in neonates (21.8% vs 5.3% in age>30 days, p=0.02) and when concomitant procedures were performed (10.6% vs 4.7% without concomitant procedures, p=0.09). No significant differences in mortality were found between index and reintervention procedures (7% vs 5%, p=0.7) or by procedure type (6% after AVR vs 5% after BAV vs 10% after SAV, p=0.5). Early mortality in the most common diagnoses was: 6%

in IAA, 11% in AVSD, 13% in HLHS, 4% in ToF, 0% in TGA-IVS, 5% in TGA-VSD, 18% in SV, 6% in DORV and 6% in PA-VSD. Early mortality decreased significantly over the study period, with an odds ratio of 0.5 per year (p=0.05), from 10.5% before 2009 to 1.2% after (p=0.01).

There were 27 neonates who required aortic intervention. Of them, 12 had an isolated AoV procedure: 9 had a BAV, two SAV (one died) and one Ross AVR. Eleven neonates underwent combined procedures: 4 Norwood operations with concomitant SAV (one died), 3 aortic arch reconstructions with SAV (one died), 2 underwent arterial switch operations (one with SAV, the other with Ross AVR) and 2 underwent combined catheter procedures (one BAV and PDA stent and one BAV preceded by an atrial septostomy). Only 4 patients in this group underwent correction of the underlying cCHD in the neonatal period prior to any AoV procedure: 3 complex aortic arch reconstructions and one arterial switch (no deaths).

***Late outcomes***

Figure 3 shows the outcomes at 10 years in all 201 patients. Neonates had the lowest survival and freedom from BAV/SAV, children had the lowest freedom from AVR but the highest survival, while adults had the highest freedom from AVR and BAV/SAV. The outcomes by age group and diagnosis are described in Tables 3 and 4, respectively.

Neonatal AoV procedures, HLHS/other SV, concomitant aortic arch repair and concomitant Fontan/Glenn were associated with increased mortality. More specifically, 5 out of 11 patients with a concomitant Fontan/Glenn died, while all 8 patients with previous or subsequent Fontan/Glenn survived. Younger age and IAA were associated with increased AVR reintervention, while smaller age, IAA and HLHS were associated with increased BAV/SAV reintervention. Procedures performed more recently were associated with lower mortality, but not with lower freedom from reintervention. Predictors of mortality and reintervention are listed in Table 5, the most important being low age, HLHS, aortic arch procedures, IAA and concomitant Fontan/Glenn.

### *Outcomes compared to patients with isolated AoV disease*

There were 133 cCHD-isolated AoV disease pairs. Early mortality in the cCHD group was higher than in isolated AoV disease (7% vs 0%, p=0.003). Patients with cCHD had lower survival at 10 years after the index AoV procedure when compared to those with isolated AoV disease patients (86.4% vs 96.1%, p=0.005, HR 4.3 for mortality). No differences in reintervention rates at 10 years were seen between cCHD and isolated AoV disease after pairing.

## DISCUSSION

Aortic valve procedures can be performed in patients with cCHD with an early mortality of 7% and late attrition concordant with the severe underlying conditions (83% survival at 10 years). More importantly, most deaths occurred in neonates and patients with single ventricle. Outside the neonatal period, early mortality in our study was lower, at 5.3%, and even lower in adults, at less than 3%. Given the severity of the conditions included, these figures should be encouraging. There was also a trend for improvement in the latter era of the study, with early mortality after 2009 being as low as 1.2%, which is in line with improvements reported in CHD surgery outcomes overall in the UK.[23]

AoV interventions in cCHD are associated with a higher early mortality (7% versus 0%) and late mortality (HR 4.3) when compared to matched isolated AoV disease patients. This excess of mortality is still in line with what we would otherwise expect, given the extreme complexity of the cases included, and can only be encouraging.

The largest cCHD cohort undergoing AoV procedures reported to date were part of a mixed AVR cohort (40% of 121 AVR patients were cCHD). They report an overall survival of 81% at 10 years, very similar to the one observed in our group, interpreted as being due to the underlying CHD.[2] Data from our matched comparison does support this notion that the natural history of complex CHD does play a major role in these outcomes, rather than the AoV procedure itself.

Of all subgroups evaluated, our study is in line with existing data on neonates undergoing AoV procedures, showing high mortality.[15, 24] All early deaths occurred when AoV intervention was required before or at the time of cCHD repair.

**Patterns by age and valvular lesion in cCHD associated with AoV disease**

We observed a trend for younger patients with AoV stenosis to have associated left heart cCHD, such as HLHS and IAA. Older patients, with AoV regurgitation, more commonly had right heart cCHD or transposition type defects. These can range from lesions in the same anatomical region, in the case of left heart cCHD, to consequences of hemodynamic changes leading to degenerative changes/annulus dilation, to incidental associations with yet undetermined causes, like those in the tetralogy spectrum. As such, they will be discussed separately below.

**cCHD of the left heart, including the aortic arch**

In cCHD affecting the left heart or aortic arch, there is an association with AoV lesions, but few cases of AoV interventions are reported.[16, 25] We found that AoV interventions in borderline HLHS are associated with increased early and late mortality, while in IAA only with increased reintervention.

The Single Ventricle Study by the Pediatric Heart Network, looking at predominantly HLHS patients, described a patent AoV in 42% of patients and moderate/severe aortic regurgitation in 1%. On the other hand, in our cohort, the HLSH patients are the “borderline” cases, where an attempt to promote left heart growth was made. Ballard et al. looked at 7 such patients undergoing hybrid Norwood in London Evelina Hospital.[9] Of those, 4 underwent AoV procedures, including one Ross operation later on. Tchervenkov also described 3 AoV procedures in 11 HLHS patients.[10] Finding that AoV procedures in these “borderline” HLHS patients are very high risk is in line with the complex nature of these uncommon, hybrid, treatment pathways, and warrants more in depth study.

Brown et al described 12 IAA patients undergoing BAV (out of 509 with congenital aortic stenosis) but did not report specific outcomes.[15] Furthermore, one out of a series of 5 patients with TGA+IAA required an AVR.[16]

**Transposition type cCHD**

In our study, patients with TGA undergoing AoV procedures were older, had valve regurgitation and similar or better outcomes than other subgroups. Fricke et al. reported that up to 2% of transposition repair patients need an AVR or SAV, which is not a negligible proportion.[11–14] Native AoV stenosis with congenitally corrected transposition is rare, described only in case reports.[26, 27]

**cCHD of the right heart and other cCHD**

The presence of AoV disease in other SV, AVSD, ToF, DORV or PA is rare and the mechanism is often unclear. We describe 106 patients with such cCHD undergoing AoV procedures of which the SV subgroup had the worst outcomes.

A previous anatomical study showed a prevalence of 1 AoV lesion in 45 ToF specimens and to date, only a few case reports are available, including two mentions of SAV.[20, 28–30] Dodds et al reported 16 patients with ToF/PA-VSD requiring AVR for aortic regurgitation, with one death.[17]

Only a few cases of AoV procedures (AVR) in SV patients are reported.[18, 19] In this group, any outflow obstruction/regurgitation can have severe implications, and the complex anatomy, both pre- and post-Fontan poses additional issues. In our study, we identified 19 patients with AoV procedures at any time during the single ventricle pathway. Interestingly, we found that concomitant Fontan and AoV procedures were associated with a very high mortality (5/11, 4 of them before discharge), while observing no deaths when the two are performed at different times.

This highlights the higher risk of Fontan procedures, but whether a staged approach is better cannot be speculated with the data at hand.

### *Limitations*

This is a retrospective study and shares all limitations specific to this methodology. Due to the procedural and retrospective nature of the dataset, the classification into cCHD, although based on consensus papers,[1, 21] is still subjective. Furthermore, limited detailed anatomy data did not allow for a better description of valve structure. A paired comparison between cCHD with and without AoV procedures could have provided extra information on this topic, but no data permitting this was available (the dataset pertains specifically to AoV procedures). The lack of clinical detail in the procedural dataset led to matching being performed based on age as a surrogate of severity, which leaves other confounders unaccounted for. The lack of data on similar cCHD patients with no AoV procedures, as well as cause of death limited the ability to differentiate the natural history of the cCHD from the impact of AoV procedures.

### *Conclusion*

AoV procedures in cCHD can be performed with good results outside early infancy, but with higher early and late mortality than in isolated AoV disease. These findings show the magnitude of the excess mortality, adding much needed evidence in a topic which was otherwise lacking in data. Neonates and patients with single ventricle defects, especially those undergoing concomitant Fontan/Glenn, have the worst outcomes. The age at intervention and valve lesion type reflect the probable underlying mechanisms and natural progression of the cCHD. These national, multicentre results can be used to aid informed counselling, adjust expectations and offer insight into several subgroups where an additional risk was observed.

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Conflicts of interest: none declared.

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## FIGURES AND LEGENDS

**Figure 1.** Age distribution at AoV procedure in the most common cCHD. Values on vertical axis represent percentage from diagnosis subgroup and those inside the chart represent number of patients for each age group and diagnosis.

**Figure 2.** AoV disease type in cCHD patients undergoing aortic intervention, by main diagnoses. Values on vertical axis represent percentage from diagnosis subgroup and those inside the chart represent number of patients for each AoV disease type and diagnosis.

**Figure 3.** Unadjusted survival and freedom from reintervention after AoV procedures in 201 patients with cCHD. Each curve represents one outcome. Values in brackets represent 95% confidence intervals. The first recorded AoV intervention represents the index procedure.

**TABLES**

**Table 1.** Demographic, clinical and procedural data in 241 aortic valve procedures in complex congenital heart disease patients

| | Total n=241 (201 pts) | BAV n=43 (33 pts) | SAV n=62 (49 pts) | AVR n=136 (119 pts) | p value |
|---|---|---|---|---|---|
| Median age, y (IQR) | 9.5 (1.5-20.8) | 0.5 (0.04-4.4) | 2.9 (0.6-10.4) | 16.8 (9.6-33.6) | <0.001 |
| Female gender (n, %) | 92 (38) | 21 (49) | 29 (47) | 42 (31) | 0.03 |
| Age group (n, %) | | | | | |
| Neonate | 25 (10) | 13 (30) | 10 (16) | 2 (1) | |
| Infant | 30 (12) | 13 (30) | 12 (19) | 5 (4) | |
| Child | 105 (44) | 14 (33) | 31 (52) | 59 (43) | <0.001 |
| Young adult (≤40y) | 59 (25) | 3 (7) | 6 (10) | 50 (37) | |
| Adult (>40y) | 22 (9) | 0 (0) | 2 (3) | 20 (15) | |
| Genetic syndrome (n, %) | 11 (5) | 3 (7) | 5 (8) | 3 (2) | 0.1 |
| Main aortic disease type (n, %) | | | | | |

| | | | | | |
|---|---|---|---|---|---|
| Regurgitation | 80 (33) | 2 (5) | 12 (19) | 66 (48) | <0.001 |
| Stenosis | 112 (47) | 41 (95) | 48 (78) | 23 (17) | |
| Unknown | 49 (20) | 0 (0) | 2 (3) | 47 (35) | |
| Procedure era (n, %) | | | | | |
| 2000-2004 | 80 (33) | 18 (42) | 30 (48) | 32 (24) | 0.002 |
| 2005-2008 | 67 (28) | 15 (35) | 11 (18) | 41 (30) | |
| 2009-2012 | 94 (39) | 10 (23) | 21 (34) | 63 (46) | |
| Follow-up, y (median, IQR) | 2.7 (0.7-7.1) | 3 (0.4-8.2) | 3 (1.7-8.3) | 2.5 (0.5-6.6) | 0.3 |

IQR, interquartile range.

**Table 2.** Complex congenital heart disease types by aortic valve procedure (n=241 procedures, 201 patients)

| | Total | BAV | SAV | AVR |
|---|---|---|---|---|
| Atrioventricular septal defect | 34 (26) | 5 (3) | 13 (11) | 16 (12) |
| Interrupted aortic arch | 34 (23) | 11 (8) | 9 (5) | 14 (10) |
| Tetralogy of Fallot | 27 (26) | 1 (1) | 6 (5) | 20 (20) |
| Hypoplastic left heart syndrome | 26 (18) | 11 (8) | 8 (5) | 7 (5) |
| TGA-IVS | 26 (25) | 0 (0) | 6 (6) | 20 (19) |
| TGA-VSD | 21 (17) | 4 (3) | 2 (1) | 15 (13) |
| Double outlet right ventricle | 19 (16) | 3 (3) | 6 (5) | 10 (8) |
| Other single ventricle | 18 (15) | 5 (4) | 5 (5) | 8 (6) |
| PA-VSD | 16 (15) | 0 (0) | 3 (2) | 13 (13) |
| Congenitally corrected TGA | 7 (7) | 0 (0) | 1 (1) | 6 (6) |
| Aortic hypoplasia | 5 (5) | 0 (0) | 1 (1) | 4 (4) |
| Ebstein disease | 4 (4) | 0 (0) | 1 (1) | 3 (3) |
| PA-IVS | 2 (2) | 2 (2) | 0 (0) | 0 (0) |

| | | | | |
|---|---|---|---|---|
| Absent pulmonary valve syndrome | 1 (1) | 0 (0) | 1 (1) | 0 (0) |
| Totally anomalous pulmonary venous connection | 1 (1) | 1 (1) | 0 (0) | 0 (0) |

Values in brackets represent number of patients.

IVS, intact ventricular septum; PA, pulmonary atresia; TGA, transposition of great arteries; VSD, ventricular septal defect.

**Table 3.** Outcomes at 10 years by age group and aortic valve procedure in 201 complex congenital heart disease patients

| | Neonates n=23 | Infants n=21 | Children n=83 | Adults ≤40y n=53 | Adults >40y n=21 | Overall n=201 |
|---|---|---|---|---|---|---|
| Survival (%) | 50 (26-69) | 85 (60-95) | 93 (80-97) | 84 (69-92) | 83 (43-96) | 83 (76-89) |
| Freedom from AVR (%) | 83 (48-96) | 93 (59-99) | 71 (52-84) | 98 (84-99) | 100 | 84 (73-91) |
| Freedom from BAV/SAV (%) | 55 (27-76) | 66 (35-84) | 92 (82-97) | 97 (82-99) | 94 (65-99) | 86 (79-91) |
| Freedom from any AoV reintervention (%) | 55 (27-76) | 66 (35-84) | 68 (50-81) | 95 (80-99) | 94 (65-99) | 75 (64-83) |

Values in brackets are 95% confidence intervals, estimates from Kaplan Meier survivor function.

AoV, aortic valve; AVR, aortic valve replacement; BAV, balloon aortic valvuloplasty; SAV, surgical aortic valvotomy.

**Table 4.** Outcomes at 10 years after AoV procedures by cCHD diagnosis type

| Main diagnosis | Deaths | AVR | BAV/SAV | Total |
|---|---|---|---|---|
| Atrioventricular septal defect | 2 | 3 | 4 | 26 |
| Interrupted aortic arch | 2 | 4 | 5 | 23 |
| Tetralogy of Fallot | 3 | 0 | 1 | 26 |
| Transposition intact septum | 4 | 2 | 0 | 25 |
| Hypoplastic left heart syndrome | 6 | 1 | 5 | 18 |
| Transposition with septal defect | 1 | 1 | 1 | 17 |
| Double outlet right ventricule | 1 | 2 | 1 | 16 |
| Other single ventricle | 4 | 2 | 1 | 15 |
| Pulmonary atresia with septal defect | 2 | 0 | 1 | 15 |
| Congenitally corrected transposition | 1 | 0 | 0 | 7 |
| Aortic hypoplasia | 0 | 0 | 0 | 5 |
| Ebstein anomaly | 0 | 0 | 0 | 4 |
| Pulmonary atresia intact septum | 0 | 0 | 0 | 2 |
| Absent pulmonary valve | 0 | 0 | 0 | 1 |
| Totally anomalous pulmonary connection | 0 | 0 | 0 | 1 |

Events include the early (30 days) period.

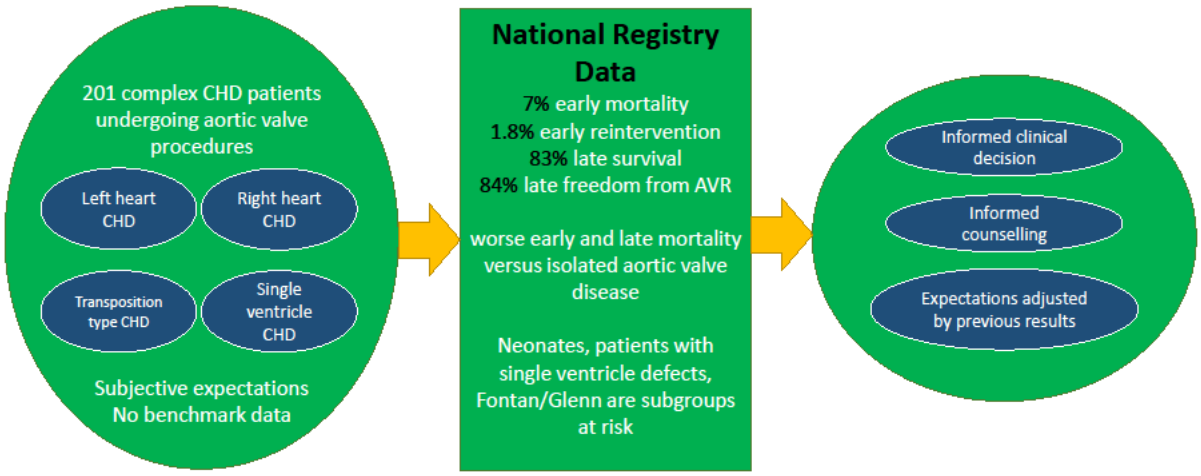
**Table 5.** Predictors of outcomes at 10 years in 201 patients with complex congenital heart disease undergoing aortic valve procedures

| | HR (95% CI) | p value |
|---|---|---|
| Mortality | | |
| Neonatal procedure | 5.3 (2.4;11.6) | <0.001 |
| Procedure year | | |
| 2000-2004 | Baseline | |
| 2005-2008 | 0.8 (0.3-1.7) | 0.5 |
| 2009-2012 | 0.1 (0.03-0.6) | 0.01 |
| Hypoplastic left heart syndrome | 3.1 (1.3-7.8) | 0.01 |
| Other single ventricle | 2.6 (0.9-7.5) | 0.08 |
| Concomitant Fontan/Glenn | | |
| vs no concomitant procedure | 6.1 (1.9-19.1) | 0.002 |
| vs other concomitant procedures | 3.5 (1.1-11) | 0.03 |
| Concomitant aortic arch procedure | | |
| vs no concomitant procedure | 4.4 (1.2-15.8) | 0.02 |

| | | |
|---|---|---|
| vs other concomitant procedures | 2.3 (0.6-8.2) | 0.2 |
| Aortic valve replacement | | |
| Age years (continuous) | 0.9/y (0.89-0.99) | 0.04 |
| IAA | 3.1 (1-9.8) | 0.05 |
| Aortic valvulopasty/valvotomy | | |
| Neonatal procedure | 7 (2.7-18) | <0.001 |
| Age years (continuous) | 0.9/y (0.8-0.9) | 0.005 |
| IAA | 3 (1.1-8.5) | 0.03 |

Results from univentricular Cox regression.

CI, confidence interval; IAA, interrupted aortic arch; HR, hazard ratio.


201 complex CHD patients undergoing aortic valve procedures
Left heart CHD
Right heart CHD
Transposition type CHD
Single ventricle CHD
Subjective expectations
No benchmark data
National Registry Data
7% early mortality
1.8% early reintervention
83% late survival
84% late freedom from AVR
worse early and late mortality versus isolated aortic valve disease
Neonates, patients with single ventricle defects, Fontan/Glenn are subgroups at risk
Informed clinical decision
Informed counselling
Expectations adjusted by previous results