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# Atrioventricular valve regurgitation in patients undergoing total cavopulmonary connection: Impact of valve morphology and underlying mechanisms on survival and reintervention

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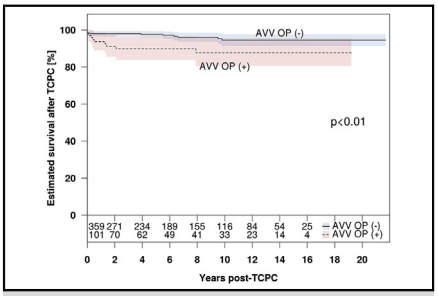
## ABSTRACT

**Objective:** The study objective was to determine the mechanisms of atrioventricular valve regurgitation in single-ventricle physiology and their influence on outcomes after total cavopulmonary connection.

**Methods:** Among 460 patients who underwent a total cavopulmonary connection, 101 (22%) had atrioventricular valve surgery before or coincident with total cavopulmonary connection.

**Results:** Atrioventricular valve morphology showed 2 separated in 33 patients, mitral in 11 patients, tricuspid in 41 patients, and common in 16 patients. Patients with a tricuspid and a common atrioventricular valve underwent atrioventricular valve surgery frequently, 27% and 36%, respectively. Atrioventricular valve regurgitation was due to 1 or more of the following mechanisms: dysplastic leaflet (62), prolapse (53), annular dilation (27), cleft (22), and chordal anomaly (14). Structural anomalies were observed in 89 patients (88%). The procedure was atrioventricular valve repair in 81 patients, atrioventricular valve closure in 16 patients, and atrioventricular valve replacement in 4 patients. Among 81 patients who underwent initial repair, repeat repair was required in 20 patients, atrioventricular valve replacement was required in 7 patients, and atrioventricular valve closure was required in 3 patients. Among patients undergoing atrioventricular valve surgery, overall survival after total cavopulmonary connection (88% vs 95% at 15 years, $P = .01$), freedom from atrioventricular valve reoperation after total cavopulmonary connection (75% vs 99% at 15 years, $P < .01$), and grade of atrioventricular valve regurgitation at a median follow-up of 6.6 years ($P < .01$) were worse than in those who did not require atrioventricular valve surgery.

**Conclusions:** Atrioventricular valve regurgitation in univentricular heart is more frequently associated with a tricuspid or a common atrioventricular valve, and structural anomalies are the primary cause. Significant atrioventricular valve regurgitation requiring surgery influences survival after total cavopulmonary connection, especially when atrioventricular valve replacement was needed. Surgical management based on mechanisms of regurgitation is mandatory. (J Thorac Cardiovasc Surg 2017;■:1-9)




Freedom from death or transplantation after TCPC in patients with and without AVV surgery.

**Central Message**

Morphologic anomalies are the primary cause of AVV regurgitation in functional single-ventricle physiology. Surgical interventions influence survival after TCPC.

**Perspective**

Our results suggest that 22% of the patients required AVV surgery before or at TCPC and that most AVV regurgitation (88%) was associated with structural valve abnormalities. Surgical intervention before or at TCPC had a negative impact on survival, AVV reoperation, and late AVV function. Although valve repair is the goal, results are not satisfactory and reoperation at the valve is high.

See Editorial Commentary page XXX.

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**Abbreviations and Acronyms**

AVV = atrioventricular valve
BCPS = bidirectional cavopulmonary shunt
CAVV = common atrioventricular valve
HLHS = hypoplastic left heart syndrome
MV = mitral valve
TCPC = total cavopulmonary connection
TEE = transesophageal echocardiography
TV = tricuspid valve

Scanning this QR code will take you to a supplemental video, figures, and tables for the article.



In patients with a functionally single ventricle, the staged Fontan palliation with a bidirectional cavopulmonary shunt (BCPS) and later a total cavopulmonary connection (TCPC) is currently a successful treatment concept. Significant atrioventricular valve (AVV) regurgitation can complicate the clinical course at any stage. In the early era of univentricular palliation, significant AVV regurgitation was a contraindication for performing the Fontan procedure.[1] However, various modifications to the Fontan procedure, improved perioperative management, and the introduction and refinement of AVV repair techniques have improved the survival of patients with a functional single ventricle and AVV regurgitation during the past few decades.[2-8] Despite such advances, recent studies have demonstrated that significant AVV regurgitation still poses a major risk for patients undergoing a staged Fontan palliation and is associated with systemic ventricular dysfunction and worse clinical outcomes.[4,6-14]

In patients with a single ventricle, AVV morphology can be complex, and the mechanisms for AVV regurgitation are often multifactorial. Various AVV repair techniques have been described, and advantages and disadvantages have been identified.[3-8,15-18] Even so, the best timing is unknown and AVV repair is technically challenging. Few studies have focused on the influence of AVV regurgitation and AVV surgery on outcomes after TCPC.[8] We investigated (1) which AVV morphology is most susceptible to AVV regurgitation and which mechanisms are involved; and (2) whether AVV regurgitation requiring surgical intervention influences survival, AVV operation rate, and ventricular and AVV function after TCPC.

## MATERIALS AND METHODS

The Institutional Review Board of the Technical University of Munich approved this study. The medical records of all 705 patients who were surgically treated for staged single-ventricle palliation at the German Heart Center Munich between 1994 and 2015 were reviewed, and 460 patients who underwent TCPC were included in this study. Because the purpose of this study was to evaluate the impact of AVV regurgitation on outcomes after TCPC, the remaining 245 patients were excluded from this study, including 32 patients who underwent AVV surgery (8 patients before stage II and 24 patients at or after stage II). AVV was classified according to the modified congenital Heart Surgery Nomenclature and Database Project classification[19]: single mitral valve (MV), single tricuspid valve (TV), 2 separate valves, and common AVV (CAVV) (single-orifice AVV associated with complete atrioventricular septal defect).

### Atrioventricular Valve Anatomy and Function

AVV and ventricular morphology, as well as structural abnormalities of the AVV, were determined from echocardiographic reports. After AVV surgery, mechanisms of AVV regurgitation were based on the surgical reports. The extent of AVV regurgitation was classified as 0 (none), 1 (trivial), 2 (mild), 3 (moderate), or 4 (severe). Moderate or severe regurgitation was considered to represent significant AVV regurgitation. Systemic ventricular function and ventricular dilatation were evaluated by echocardiography. Ventricular function was graded by the investigator as normal (ejection fraction >50%), mildly reduced, moderately reduced, or severely reduced.

### Indication and Timing of Atrioventricular Valve Surgery

Moderate AVV regurgitation was added to the staged surgery, and severe AVV regurgitation was addressed in a separate procedure between the staged procedures. Indications for AVV surgery were based on transthoracic echocardiography findings and confirmed by intraoperative transesophageal echocardiography (TEE). Usually, severe recurrent AVV regurgitation was treated as soon as it was detected, whereas moderate recurrent AVV regurgitation was addressed at the next stage of palliation.

### Operative Techniques

AVV repair was performed under standard cardiopulmonary bypass with cardioplegic cardiac arrest. Surgical techniques were individualized according to the valve pathology reported from the echocardiographer. Intraoperatively, saline was injected into the ventricle to confirm the mechanism of AVV regurgitation. Attention was paid to the annular dimension, commissural leak, prolapse or restriction of the leaflets, and leaflet and subvalvular abnormalities. Local annuloplasty and commissuroplasty were the primary techniques for treating annular dilatation, leaflet prolapse, or both (Video 1).

Different types of annuloplasty were performed: usually, a 5-0 or 6-0 Prolene horizontal mattress suture was placed along the commissures where the predominant regurgitant jet was detected. Partial annuloplasty was performed in patients with prolapse of 1 or 2 leaflets, resulting in failure of coaptation. Circular annuloplasty was performed using the De Vega technique. An annuloplasty ring was used only in patients with an adult-size annulus.

Commissuroplasty was achieved with edge-to-edge approximation of the leaflet tissue using 6-0 or 7-0 Prolene sutures. Occasionally, the posterior leaflet of the TV was obliterated using the method of Ohye and colleagues.[4] If localized or generalized prolapse occurred when the chordae and subvalvular apparatus of the adjacent leaflet were essentially normal, this was treated with edge-to-edge approximation of the commissures to attach the prolapsing region to the nonprolapsing region.

In patients with poor coaptation of 2 opposing leaflets, suturing of the free-floating segments of the opposing leaflets created double orifices, and edge-to-edge repair was performed. If a single cleft or dehiscence was the primary cause of regurgitation, it was primarily closed with interrupted Prolene sutures. If the valve leaflets were dysplastic and had

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