Right Heart Failure in Pediatric and Congenital Cardiac Surgery

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Introduction

The Morphologic Right Ventricle in Subpulmonary or Systemic Position

There cannot be too much argument about the notion, that the right heart, in a way or another, is involved in the majority of structural congenital heart defects. For a long time, however, the right ventricle as a subpulmonary ventricle has been regarded merely as an appendix to the systemic, mostly left ventricle, which from an embryological view actually is completely true [82]. This belief, that the pulmonary circulation, also known as lesser circulation is of lesser importance and of an undemanding nature, was further supported by the fact that patients can attain a near normal lifestyle during childhood after a Fontan operation: An operation where the circulation is completely devoid of an immediate subpulmonary ventricle.

With time the function of the right ventricle in particular, and the subpulmonary ventricle in a broader sense emerged as issues of serious concern when patients thought to be healed by corrective surgery early in live reached maturity: a plethora of negative events occurred. It was discovered almost simultaneously that the dysfunctional right ventricle, whether it represents a systemic pump or is chronically volume overloaded in the subpulmonary position post surgery deserves more attention than previously thought. The numerous detrimental issues arising in the ageing and continuously growing post-Fontan population further substantiated the newly awakened interest in the physiologic and pathophysiologic parameters of the lesser circulation. These causes and sequelae clinicians have been unaware of before.

It was the group at the Royal Brompton and National Heart and Lung Hospital London, England, led by Andrew Redington who first questioned the benign nature of the large right ventricle following TOF repair [10, 31]. At the opposite side of the globe, encouraged by the success of the arterial switch operation as a therapy for transposition of the great arteries, the surgical group from Royal Children's Hospital

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Melbourne, Australia, under the leadership of Roger Mee [58] had begun earlier to revert patients with failing systemic right ventricles years after atrial switch surgery. This resulted in anatomically corrected normal, concordant ventriculo-arterial connection.

A complete and comprehensive discussion of the issue of a failing (or lacking) right ventricle comprises many different aspects and a structured, logical inventory is rather complex. We will distinguish between two settings with two subsettings each. The first setting is the inadequacy of the cavopulmonary pathway which we consider as the clinical syndrome of right heart failure, it may occur with or without subpulmonary ventricle, in a bi- or univentricular subsetting. The second setting is the failure of the morphological right ventricle in the systemic circulation, also with a bi-or univentricular subsetting.

The Clinical Syndrome of Right Heart Failure: Inadequacy of the Cavo-pulmonary Pathway with or without Subpulmonary Ventricle

For many readers the term cavo-pulmonary pathway is clearly associated with the Fontan type of circulation. In the following, however, we consider its inadequacy as a clinical entity characterized by insufficient return of the systemic venous blood into the pulmonary circulation, independent from the presence or absence of a subpulmonary ventricle, almost always a right ventricle.

The classical clinical signs of peripheral venous congestion with interstitial and intracavitary water retention are the hallmarks of this syndrome. If communications at atrial and/or ventricular level are present peripheral desaturation (cyanosis) can also be observed. Currently the full-blown chronic clinical syndrome is rarely seen in the pediatric age group due to rigorous medical and surgical follow-up of patients with operated or non-operated congenital heart disease. There is, however, also an incidence of acute early postoperative failure.

The issues arising from inadaequate systemic venous to pulmonary pathway, encountered as sequelae of congenital heart disease, can be categorized as being either acute or chronic.

Acute issues:

- acute postoperative right ventricular failure
- acute dysfunction of a systemic-pulmonary shunt
- neonates with congenital heart disease:
  - partially or completely ductal dependent pulmonary circulation
  - rare conditions with non ductal dependent pulmonary circulation

Chronic issues:

- a failing Fontan circulation
- preoperative decisions regarding incorporation of "small" right ventricles into the pulmonary circulation
- late sequelae of the right ventricle after previous surgery

The issue of Eisenmenger syndrome was intently omitted being considered a more or less historical and fortunately vanishing entity.

Acute Issues

Acute Postoperative Right Ventricular Failure

The right ventricle may fail acutely in the early postoperative hours or days due to many potentially reversible intraoperative issues such as inadequate myocardial protection, large incisions on the free wall or extensive septal reconstructions and last but not least pulmonary hypertensive crises.

Pulmonary hypertensive crises, once dreaded complications of corrective surgery of left to right shunt lesions performed beyond infancy, are currently less frequently encountered and generally have a limited clinical impact. This positive trend is due to the fact that now a days timing of the operations is well before the development of increased pulmonary resistance.

There is also the current awareness of the caring personnel and the preventive measures available in standardized postoperative care. Despite these measures, however, dramatic forms of acute pulmonary hypertensive crisis may be encountered, especially after repair of an obstructed total anomalous pulmonary connection (in simple and complex heterotaxy forms either) and in infants with trisomy 21 whose pulmonary vascular tone is particularly unstable and unpredictable, especially when repair is performed late in infancy. The condition is reversible and the success of recovery is highly dependent upon a prompt management, which should include establishing immediate extacorporeal life support after 10 min of CPR in patients not responding to pharmacologic support [25].

The preferred support modality for any acute severe cardiac or pulmonary dysfunction is a veno-arterial extracorporeal membrane oxygenation (ECMO) setup. This is functionally a standard cardiopulmonary bypass circuit with extracorporal circulation as employed in any open heart surgery these days. The use of the PediMag (modified CentriMag) circuit marketed by Thoratec Corp (Pleasanton, CA) has made setting up and maintenance of ECMO a routine. In the early postoperative patient the circuit is established between the right atrium and the ascending aorta, although cannulation of the neck vessels in the young, as well as the groin or iliac vessels in the occasional older patient could be considered. The latter especially, if ongoing sufficient mechanical chest compression outside the operating room, is prevailing. Lack of an adequate aortic cannulation site may prevent intrathoracal cannulation also.

In modern pediatric ECMO systems centrifugal pumps and heparin coated hollow fiber membrane oxygenators have replaced the roller pumps and silicone membrane oxygenators used earlier. Heparin coating of the entire circuit, requiring less anticoagulation have greatly improved the management of time and resources consuming bleeding complications encountered earlier. Full Heparinisation, however, has still to be considered when the ventricle(s) is (are) not ejecting. The support of the neonates with shunt dependent univentricular circulation posed some controversies in the past concerning shunt management. The previously recommended temporary shunt closure during support has been circumvented by doubling, or at least increasing the flow of the assist device. The temporary shunt closure entails the risk of thrombosis, so that shunt revision is eventually mandatory during ECMO weaning. The high flow rates compensate for the pulmonary run-off and offer a more physiologic environment for the pulmonary circulation. Should the pulmonary function be of no concern early postoperatively one can perform the assist without the inclusion of an oxygenator. This concept known as the NOMO VAD (no membrane oxygenator ventricular assist device as in contrast to ECMO: extracorporal membrane oxygenation) was popularized by Ross Ungerleider and his group [15], the authors recommending it to be routinely performed during the first 24–48 h postoperatively in patients undergoing stage I. reconstruction for hypoplastic left heart syndrome.

Results of ECMO support for reversible right heart dysfunction are encouraging. Outcome analysis of support instituted intraoperatively due to inability to wean from CPB after open heart surgery remain poorer than those instituted after a period of relative stability or acute postoperative failure. The database of the ELSO (Extracorporal Life Support Organization) for patients suffering from congenital heart disease has been reviewed many times since its establishment in 1986. The benchmark outcome data of 60 % successful weaning and 40 % successful hospital discharge [26] are only marginally improving over the time, these results being confirmed by numerous single institutional reviews, also. The different efforts to identify outcome predictors led to very conflicting results, so that it may be postulated that the lack of predictive factors makes every patient a potential candidate for ECLS (ECMO, NOMOVAD).

Acute Dysfunction of an Aorto Pulmonary Shunt

Acute shunt dysfunction in a palliated neonate and infant is almost always a fatal event. Besides tech-

nical issues which usually occur early postoperatively [64] dehydration and infection represent the main causes of shunt thrombosis. With rare exceptions the establishment of rapid circulatory support represent the only therapeutic modality in the event of an acute hypoxemic circulatory collapse. As highlighted earlier in neonates and infants outside the early preoperative phase the peripheral cannulation is performed via the rightsided neck vessels.

Neonatal CHD with Non-ductal Dependent Pulmonary Circulation

Due to the persistence of functional intrauterine shunts many congenital heart defects with potentially ductal dependent pulmonary circulation rarely manifest themselves as an acute insufficiency of the pulmonary circulation immediately after birth. Gradual ductal closure within the first days heralded by deepening cyanosis alerts the neonatologists. Reviewing the abundance of conditions with completely or partially ductal dependent pulmonary circulation is beyond the scope of this chapter, yet some conditions with borderline right ventricles present at birth will be dealt with in the following subchapter.

Nevertheless there are some rare conditions with severe cyanosis present right after birth where the physiologic closure of the ductus arteriosus is not involved in the newly developing pathomechanism.

Acute intrauterine tricuspidal insufficiency produced by papillary head avulsion due to acute and massive RV afterload increase secondary to premature closure of the ductus arteriosus [50] or a congenital large PA to LA fistula [60, 85] are some of the conditions which are relatively straightforward to treat but represent an extreme perinatal emergency. Success can be only expected after immediate postpartal diagnosis, or better: fetal diagnosis.

Acute/Chronic Issues

The Small Subpulmonary Ventricle and the One-and-a-Half-Ventricle Concept

The previous chapter has highlighted the benefits of incorporating a subpulmonary ventricle in the cavopulmonary pathway, also as a mechanical device. It is therefore mandatory to consider incorporating an even distinctly small pumping chamber between the systemic veins and the pulmonary artery in any setting of complex congenital heart disease. The commitment to a high risk septation to reach a complete biventricular repair in anatomically equivocal hearts may be more disadvantageous, as large intraventricular baffles and tunnels might not only lead to an inadequate RV volume but can also compromise ventricular outflow [21]. Yet there are some well-described anatomical entities where a borderline ventricle can handle the IVC return, with the SVC being directly connected to the pulmonary arteries.

This concept originated in the early 1990s [34, 80] and is routinely used in hypoplastic right hearts such as left dominant imbalanced AV-canals/AV-septal defects, Ebstein's anomaly and Pulmonary Atresia with Intact Ventricular Septum (PA/IVS).

Left Dominant AV-Canals/AV Septal Defects

This anatomic variant is significantly less frequently encountered in patients without heterotaxy than the right dominant form, yet the criteria of Cohen (1997) with its more recent modifications [46] are helpful to distinguish them preoperatively. A staged reconstruction with primary pulmonary artery banding followed by second stage intracardiac repair and a superior bidirectional cavo-pulmonary connection (BCPC) is the currently preferred strategy for these patients.

As a bail-out intraoperative maneuver in the presence of a preoperatively unprotected pulmonary arteries in a baby with reactively increased pulmonary vascular resistance the unloading of the hypoplastic right ventricle by an adjunctive bidirectional cavopulmonary connection is not a safe maneuver, as the elevated pulmonary resistances might prohibitively compromise the outflow from the SVC.

In cases where a previously unrecognized RV hypoplasia presents as an acute intraoperative problem after an already completed intracardiac biventricular repair, a temporary systemic-pulmonary shunt, with atrial septal patch fenestration (alternatively ventricular septal patch

fenestration) is sometimes the only alternative to a total takedown of the intracardiac repair. A BCPC with closure of the previously created left-right communication (s) can be performed after documented normalization of the pulmonary vascular resistance.

In the rare patient with low pulmonary resistances the BCPC is performed straightforwardly during the operation for AV canal repair.

Ebstein's Anomaly of the Tricuspid Valve

Besides the extreme neonatal form, which requires immediate, or even emergency attention, these patients will become symptomatic later in life. The leading clinical symptoms presenting are very useful for the selection of patients with inadequate RV. The presence of severe preoperative desaturation in the presence of an ASD may be also a sign of the incapacity of the right ventricle to handle the entire cardiac output and the plan for the one and a half ventricle repair has to be considered. The different aspects of the repair of an insufficient Ebstein valve [9, 14, 16, 41] are beyond the scope of the current chapter, yet the desire and efforts to obtain a competent tricuspid valve might end up in creating a functionally stenotic tricuspid orifice. Given the poor prognosis of a prosthetic valve in the tricuspid position in small children, a restrictive but competent tricuspid valve, especially in pediatric patients is a better alternative than a normal sized prosthesis. Under these circumstances the solution is also a BCPC. Malhootra et al. [53] in a recently published very elegant paper delineate an intraoperative decision algorithm, where the indication for the need of an unloading BCPC can be decided upon the intraoperative postrepair hemodynamics. Any postrepair RA:LA pressure ratio above 1.5 should prompt a one-and-a-half repair.

Pulmonary Atresia and Intact Ventricular Septum (PA/IVS)

The issues discussed for Ebstein's anomaly can generally be applied for PA/IVS as well, except that in these patients a neonatal stage I palliation is mandatory. Extremely underdeveloped right ventricles with lacking inlet or outlet components [18], will clearly speak against any attempt of recruiting the right ventricle. In patients where the neonatal size and morphology of the right ventricle is ambiguous opening the right ventricular outflow tract by a transanular patch completed by a systemic-pulmonary shunt could stimulate "catch-up growth" of the hypoplastic right ventricle.

The decision for the further reconstruction strategy to be pursued should be taken at the time of the second stage operation usually around 6 months of age. Despite some isolated reports upon growing right ventricles following opening the RVOT by inserting a transanular patch in the neonatal period [28, 73] the majority of the literature data do not support the idea of size gain in ventricles where the initial Z- value of the tricuspid valve was under −4 [39]. Patients for whom an eventual one-and-a-half ventricle reconstruction can be seriously contemplated are those with initial tricuspid annular Z values between −4 and −2. They represent a minority among the patients with PA/IVS, their proportion being about 5 % of all patients born with PA/IVS [2].

Chronic Issues

The Sequelae Right Ventricle, the RV after RVOT Surgery

The right ventricle tolerates the deleterious hemodynamic consequences of valve related volume overload for a relatively long time after corrective surgeries in infancy and early childhood. Occasionally dysfunctional valves may result from avoidable technical imperfections, but in their majority they represent a clear choice taken by the surgeon. The perfect case for this "lesser of two evils" compromise is the transannular patching (TAP) and/or pulmonary valvectomy during right ventricular outflow tract reconstruction in patients presenting with Tetralogy of Fallot (TOF). Classical repair in the 1960s and 1970s consisted of an extremely large incision extending into the body of the right ventricle far beyond the lower boundaries of the infundibulum. The length of the incision combined with the extent of the resection of the hypertrophied wall as well as a sometimes very generous (wide!) transannular patch size left the patients with a more or less free regurgitation from the pulmo-

nary arteries into an surgically impaired right ventricle.

Due to the compliant nature of the pulmonary arteries in childhood the volume overload initially is handled well by the hypertrophied and less compliant right ventricle. The progressive regression of the hypertrophy as well as ageing of the pulmonary arteries causes progressive ventricular dilatation in large number of patients. Gatzoulis in a series of benchmark studies demonstrated that an end diastolic RV volume of 180 ml/m^2 as well as a QRS length of >160 ms can be considered cut-off values, above which the incidence of malignant ventricular arrhythmias as well as the contractile dysfunction is significantly higher [32]. These data were further supported by the results of other investigators [45, 74].

Limiting the length or complete avoidance of the infundibulotomy, or even ventriculotomy, and performing only minimal or completely avoiding muscle resections are considered by many cornerstones of an efficient preemptive strategy [3, 45, 63]. Others recommend so called pulmonary valve sparing, but rather function preserving, reconstructive strategies [1, 76] and the implantation of an autologous pericardial monocusp valve during the initial surgery [38].

Whether these recent modifications will stand the trial of time regarding their efficiency to reduce the incidence and extent of the pathology related to the unguarded right ventricular outflow tract is not clear yet. There are clear indices that the magnitude of regurgitation, ventricular dilatation and dysfunction following TAP repair are more profound, but patients who have undergone transatrial repair are presenting with progressive dilatation, also [7, 24, 81, own not published experience]. Anatomically the pulmonary valve, devoid of a proper fibrous annulus is suspended inside a muscular collar represented by the infundibulum. As the infundibulum is characteristically involved in the obstruction seen in TOF, surgical manipulation will always alter its architecture. Even initially well-developed and unobstructive pulmonary valves will have an altered and weakened muscular support prone to dilatation and ultimately leading to valvular insufficiency.

Moreover there are claims that the infundibulum itself is having a sphincter like function, which is profoundly altered in TOF patients.

TOF patients are the most numerous but not the only category where a valveless communication is established between the right ventricle and the pulmonary arteries following corrective surgery. In a large, homogenous and carefully documented follow-up series [22] of the valveless REV procedure ("reconstruction a l'etage ventriculaire") applied for TGA/VSD/PS after an average of 12 years the need for secondary pulmonary valve implantation was strikingly lower 5/171 (2.7 %) as it would have been expected in a similar TOF population. RVOT-Reoperations in a European Multicenter Study, however, was as high as 40 % [40].

Addressing right ventricular systolic and diastolic function in patients following biventricular PA/IVS repair in adolescents Liang and coworkers [52] found surprisingly high incidence of a restrictive right ventricle (81 %) with low volume indexes and almost normal exercise capacity. Other reports on this patient group found a significantly higher proportion of dysfunctional tricuspid valves altering RV function than in the control group of TOF patients, which the authors ascribe to the dysplastic nature of the tricuspid valve in patients with PA/IVS [5]. Although consistent data on long term outcome of these patients are lacking, in the light of the above findings the incidence of reoperations on the RVOT will be eventually lower but this will be biased by a higher incidence of tricuspid valve interventions.

Pulmonary Valve Implantation

After repair of TOF and other similar conditions with eventual RV volume overload, the majority of the adolescents are completely asymptomatic. With changing echocardiographic and MR indexes, the indications for intervention are not clear-cut for the time being. Improvements of the RV systolic function and reversal of the RV dilatation have been documented by some reports, but these findings were not universally supported. From the multitude of studies whose methodologies and results significantly differ

from each other, it is hard to exactly formulate values on which a pulmonary valve implantation should be decided upon. Age ≥17.5 years [30], altered LV function [35] and end diastolic volume indexes above 170 ml/m^2 (Therrien 2005) are among the many other predictors found accountable for an unfavourable outcome following pulmonary valve implantation. During recent years the end diastolic RV volume indexes where an operation should be contemplated came down progressively from 200 to 150 ml/m^2 BSA [30] or even lower. Given the extreme reduced mortality and morbidity of this intervention this policy does not seem exaggerated. Simple infundibular valve implantation in form of a homograft or other biological valved conduits suffice for the moderately dilated ventricles. Larger ventricles, especially in patients operated upon in earlier eras by a wide and long transannular patch reaching deep into the ventricle, the thinned out non functional anterior wall after previous resection may have to be excised to improve the mechanical efficacy of the ventricle. This operation which factually is equivalent to a postinfarction left ventricular aneurysmectomy, is called RV remodeling and is gaining more and more acceptance in the recent years [20, 37]. The first midterm results of such an aggressive approach could not show a benefit over pulmonary valve replacement alone [36].

Conclusions: In a recent review on the topic Kantor and Redington [47] concluded, that: "some lesions (single-ventricle physiology, tetralogy of Fallot, and systemic RV amongst others) are never completely repaired, and others may have persistent abnormal hemodynamic loading abnormalities; patients with these remain at risk of postoperative heart failure. Treatment is multifaceted and relies on focused surgical reintervention, timely medical therapy, and, occasionally, innovative measures, such as resynchronization therapy". We can only humbly agree with them…

Our aggressive pulmonary valve replacement at RV-enddiastolic volumes between 120 and 130 ml cc/m^2 had led to 40 % cumulative valves implanted, in a group of 116 consecutive primary infant and neonatal TOF repair after 12 years (Unpublished data). At this time there was no difference between transatrial and transinfundibular repair. A final benefit of better preserved RV-function or the disadvantage of additional interventions may be recognized only after 20–30 years.

The failing Circulation Late after Fontan-Kreutzer Surgery

The Fontan-Kreutzer operation has in many ways changed not only the fate of children born with heart defects unsuitable for biventricular repair, but also the way clinicians perceive hemyodynamics, particularly the hemodynamics of the pulmonary circulation. The clinical experience has confirmed what earlier experimental studies have by then demonstrated, namely that the entire pulmonary blood flow can be propelled at the expense of only 6 mmHg pressure drop. Yet it lasted almost 20 years until the maturation of the strategy made it possible to reach the current operative mortality of around 5 % [49, 56, 75] with a relatively low postoperative attrition rate in the first 20 years of life. The stepwise improvement of patient selection, introduction of the staged reconstruction, as well as the many technical refinements helped to prevent many of the complications seen earlier, nevertheless skepticism concerning the long term fate of the Fontan operation, or rather the fear of imminent failure in the adulthood persists. This fear [43] is based on two assumptions: first the ageing process will stiffen the pulmonary vessels determining an increase in the pulmonary vascular resistance, and second the age determined reduction of the ventricular compliance will lead to the increase of the ventricular preload. This ventricular preload in a Fontan-Kreutzer Circulation had been relatively low before leading to chronic underfilling of the systemic ventricle and therefore finally adding to ventricular failure.

All of these factors, tolerated and also easily adjusted in a biventricular setting, will increase the impedance the systemic and pulmonary flow has to work against, and will ultimately cause a progressive hemodynamic alteration ending in circulatory failure.

In lack of concrete clinical experience it is hard to predict at which age the Fontan-Kreutzer circu-

lation will ultimately fail, educated pessimists anticipate it to happen in the fourth decade [72].

During the recent years many attempts have been made to develope a therapeutic modality for these patients. Optimization of the hydrodynamics [19, 62] by converting the less efficient atriopulmonary connections to a more efficient cavopulmonary pathway with eventual complete exclusion of the atria from the Fontan circuit [55] and aggressive management of atrial arrhytmias [17] represent all well justified and beneficial interventions, yet the ultimate therapeutic modality of a cardiac transplantation or some form of long term mechanical circulatory support should always be kept in mind as the Fontan population becomes older.

As long-term mechanical support for left ventricular failure has become an accepted and viable alternative for many patients suffering from acquired end stage heart disease the perspective of supporting these patients will become a definitive challenge for specialists treating these patients in the next few years.

The precise aims of the support need also to be defined: Should it be a temporary support or a rather a destination therapy? Should the device assist the ventricle or should it be limited to the cavopulmonary pathway? These are just few of the many questions the current clinical experience cannot answer yet, and which need to be cleared relatively soon [69].

Transplant policies across the world are more and more strictly regulated. It is not clear how patients with relatively well preserved ventricular function, but a prohibitively elevated CVP will fit into the urgency listings prior to development of a more or less irreversible kidney or liver damage. Although the liver and kidney function of supported patients recovers relatively promptly [67] under isolated cavopulmonary mechanical support, so that theoretically a temporary withdrawal can be contemplated if the systemic ventricle shows an acceptable function, the benefits of such a policy are more than questionable since the pulmonary vascular and diastolic properties of the ventricle are unlikely to reverse during the support phase. It is therefore more realistic to view the cavopulmonary support as a destination or "bridge to transplantation" rather than a "bridge to recovery" therapeutic modality. Certain design requirements have been postulated during the recent years, two of them being particularly important to consider:

- the need for an adequate inflow chamber
- the reduced passive internal flow resistance of the device in case of pump failure.

The currently available axial flow pumps are adequate for a cavopulmonary support, but adaptations are definitively required. As the cavopulmonary pathway has a low pressure and energy demand, the current axial pumps need to be modified in terms of pressure/suction relationship. From an engineering standpoint, at least in computer models, this is completely feasible [65, 78, 83]. Some of these devices have already been materialized beyond the stages of computer-assisted design, but there are not yet in the phase of animal studies or clinical trials.

The proposed anatomical approaches for the implantation are also different. While the group from the University of Colorado is working on a long-term support device, implanted surgically, others favor percutaneous devices.

Both approaches have their benefits and shortcomings.

The first approach needs an extended surgery to reconnect the two caval veins and redirect the blood into the pulmonary artery through an interposed segment containing the axial flow pump [84]. This variant is certainly more efficient since it creates effectively a biventricular circulation. The objections against this type of device include the need for an extensive surgery and the definitive/irreversible character, which does not allow explantation and reversal to a Fontan circulation without further major surgery. The other approaches favor percutaneously introduced expandable devices, which are meant to augment solely the IVC return. Such a system is rapidly deployable without any need to alter the existing pathways, and the model worked on by the group from Indiana University [69] is even claimed to be able to decompress the SVC, too, using a pump based on Kármán's principles [68]. Time

will tell which system will prevail, but in our opinion, the two modalities should be regarded rather as complementary: the role of the percutaneous systems being the "bridge to bridge", while the surgical systems will become the modality for longer term support. Whether the percutaneous "4-way" device, once available for neonatal or infant use will revolutionize surgery for leading to a neonatal Fontan-Kreutzer Procedure with temporal two ventricle dynamics has to be speculated. Also speculation is the potential to ameliorate the current longterm problem.

There are no published clinical or experimental attempts of using intracorporeal pneumatic devices, yet successful survival to transplant with an extracorporeal pneumatic pump is documented [67].

Ventricular dysfunction is the other failure modality of the Fontan patients. From a theoretical standpoint they can be regarded as suffering from biventricular failure, but the need for a biventricular support is questionable. Univentricular support devices have been successfully implanted as rescue "bridge to transplant" strategies for perioperative failure [29, 63, 71], but results of a systematic program to address a progressive ventricular decline in Fontan patients has not been reported, yet.

The Morphological Right Ventricle in the Systemic Circulation

Biventricular Setting: Congenitally Corrected Transposition of the Great Arteries (ccTGA)

Transposition complexes are defined as congenital heart defects with ventriculoarterial discordance with or without additional atrioventricular discordance. In a broader sense cases of univentricular hearts with right ventricular morphology can be included in this group as well.

Although the first arterial Switch operation was published 1975 [Jatene], functional repair of complete transposition remained to the late 1980s. Switching the circulation at the atrial level, the morphologically RV ended up supporting the systemic circulation. The subpulmonary left ventricle rapidly thins out and developes a crescent like shape in this configuration, while the right ventricle becomes circular in cross section and its wall thickness is adapting to the systemic requirements.

The introduction of the arterial switch operation not only changed the treatment strategy for transposition of the great arteries, but also highlighted some of the intimate details of the dynamics of the many adaptative mechanisms which take place not only at macroarchitctural but also at histological and cellular level when switching a ventricle form the systemic to the pulmonary circulation and vice versa [11, 23]. The bullet-like elliptical shape of the morphologically left ventricle makes it a more efficient high pressure pumping chamber as opposed to the L-shaped right ventricle. The relatively convergent alignment of the subvalvular elements of the mitral valve as opposed to the more divergent disposition of the tricuspid valve predestines the former to better handle not only pressure but also of volume loads without becoming insufficient [82].

Subtle subclinical changes [42] as well as reduced exercise capacity are documented in the majority of the patients with systemic right ventricles. In addition to the issues related to the systemic right ventricle and the systemic tricuspid valve these patients experience various problems later on due to obstructions in the atrial baffles and rhythm disturbances.

Patients after an atrial switch operation performed during infancy and early childhood are relatively free from ventricular dysfunction, and experience normal life during their first two decades. Longitudinal studies, however, demonstrate a progressive decline of the clinical state much later after the operation [70].

In contrast to the patients with an intact ventricular septum those who presented originally with a VSD as part of their anatomy have a significantly worse outcome in both the s/p atrial switch (d-TGA) and the operated upon congenitally corrected Transposition (ccTGA) patients. Both subgroups with VSD show a higher incidence of tricuspid insufficiency.

Whereas some ccTGA patients can present with dysplastic, Ebstein-like tricuspid valve from

birth, the mechanism implicated in tricuspid insufficiency in systemic right ventricles seen postoperatively is almost always secondary to leaflet distortion induced by the placement of the VSD patch. In addition septal dysfunction secondary to patch suture placement also may play a role [27]. In a recent report Szymanski et al. [77] studied the mechanism of tricuspid insufficiency in systemic right ventricles. They demonstrated in the majority of their patients the typical tenting, characteristic for functional mitral insufficiency seen in left ventricular dysfunction: downward tethering of the leaflets correlating well with the dilatation and systolic dysfunction of the systemic RV.

In a downward spiral, this secondary RV volume overload with the divergent arrangement of the tricuspid subvalvar apparatus present, further tethers the tricuspid leaflets impeding the valvular function and further worsening the ventricular dysfunction.

A secondary arterial switch as treatment option for these patients was inaugurated by the group of Roger Mee [58]. Their left ventricular retraining was achieved by placing a pulmonary artery band based on an earlier technique introduced by Yacoub [79]. The strategy of preliminary banding, although a relatively straightforward procedure seems to play an important role of the ultimate success of the operation. The group of Mee more and more moved to a stepwise tightening to avoid primary LV dysfunction, preferring 1.5 reoperations/patient, whereas Brawn's group in Birmingham prefers a one stage banding to the LV:RV target ratios ranging between 65 and 80 %. As technical variants to the progressive banding some groups use off-label adjustable gastric bands [6], but a bulky dedicated telemetry driven system (Flow-Watch) [12] is also available. Independent from the strategy used to prepare the LV, late dysfunction of the left ventricle seems to develop in some of the patients following this "anatomic repair" [4, 8] so that many groups are introducing novel policies. One of them consists of routine PAB in even asymptomatic neonates born with ccTGA [13, 59]. The advantage of such a routine banding is that the LV is switchable at any age, should the patient become symptomatic, and additionally the septal shift preserves the RV geometry and theoretically prevents a secondary subvalvular left ventricular obstruction. Recent animal studies [51, 61] suggested a more physiologic LV training by applying a chronic intermittent (12 h on/12 h off) banding, the function of the LV being superior to the continuous controls. Patients in the ccTGA group are often presenting with subvalvular or valvular stenoses in the left ventricular outflow tract, which can interfere with the arterial switch. In these patients a Rastelli type of reconstruction was added to an atrial switch. As the aortic root often is relatively remote to the VSD, the intraventricular Rastelli tunnel can be very voluminous. This is further aggravated by the fact that resection along the anterocephalad rim of the VSD, bearing the conduction tissue cannot be performed. The direct consequence of large intraventricular baffles is reduction of the right ventricular volume. These problems were addressed in two different ways.

The group from Stanford [54] liberally performs a BCPC to unload the right ventricle in this setting. Doing this, an inferior Hemi-Mustard procedure, completes the double switch procedure, which in their view promises less long term atrial rhythm disturbances and baffle obstructions. They deem this approach especially advantageous in hearts with atrio apical discordance where the pulmonary venous atrium is very diminuative, which further complicates atrial reconstruction.

Another method to circumvene this issue is the performance of a Nikaidoh type aortic translocation, which is also very advantageous in the prospect of later subaortic obstructions along the ventricular baffle [44].

From the accumulated experience of numerous groups pursuing a strategy to retrain the LV and obtain an anatomic correction for a dysfunctional systemic RV some fundamental principles can be derived.

– the indication for a switch-back strategy and retraining the LV should be put early, as soon as significant tricuspid dysfunction occurs.

- the need to achieve at least a two-thirds systemic pressure without significant LV dysfunction prior to the secondary arterial switch.
- the need for careful monitoring of the cardiac chamber shape and of the hemodynamic parameters to recognize the requirement for secondary band adjustments.
- the ability to retrain the left ventricle is progressively lost with increasing age, and the time required therefore maybe too long, so that currently patients beyond adolescent age are not candidates for an LV retraining strategy, anymore.
- even in patients where the ultimate goal of incorporating the LV in the systemic circulation cannot be obtained, the pulmonary artery banding is beneficial as it shifts the septum back towards the right ventricle and thereby may ameliorate the functional component of the tricuspid insufficiency;
- in patients where a switch back strategy is improbable, early aggressive treatment of the tricuspid valve insufficiency is recommended.

The Right Ventricular Dependent Univentricular Circulation

In univentricular hearts where a morphologically right ventricle acts as a systemic pump it was intuitively anticipated, that it would perform very poorly and the rate of midterm attrition due to ventricular failure would be significant. Yet the clinical experience with the repair of HLHS and other similar univentricular defects has not confirmed this presumption [33, 57] so far.

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