A Review of the Management of Pulmonary Atresia, Ventricular Septal Defect, and Major Aortopulmonary Collateral Arteries

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Background. **The management of pulmonary atresia with ventricular septal defect (PA/VSD) and major aortopulmonary collateral arteries (MAPCAs) has significantly changed over the past 20 years. Unifocalization and rehabilitation have been described as diametrically opposed strategies. An updated review focused on the management of this complex and rare condition is needed.**

Methods. **Articles related to PA/VSD/MAPCAs issued until December 2017 were screened. Twelve main studies published in the modern era (since 2000) were selected and analyzed.**

Results. **Unifocalization and rehabilitation respectively focus on the mobilization of collateral arteries and the growth of native pulmonary vessels. A third strategy, called "combined strategy," was distinguished from the review of the literature. Surgical cohorts and methods of data reporting were found to be heterogenous. Outcomes, regardless of the strategy, have transformed the natural history of the condition, with a complete repair rate of approximately 80% and low rates of early and late mortality. Patients with the most unfavorable anatomy (absent central pulmonary arteries and hypoplastic MAPCAs) remain a challenge and are still left palliated.**

Conclusions. **Variable surgical strategies are used in the management of PA/VSD/MAPCAs. Most teams report a repair rate of 70% to 80% and a mortality rate lower than 10%. Standardization in data reporting is necessary to better compare the existing strategies.**

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Pulmonary atresia with ventricular septal defect (PA/VSD) is the extreme form of the spectrum of tetralogy of Fallot (TOF) [1], with an incidence of 0.07 per 1,000 live births that accounts for approximately 2.5% of all congenital heart diseases. The majority of cases have a normally developed pulmonary blood supply, but 20% to 40% of cases will have multifocal blood supply to the lungs—through major aortopulmonary collateral arteries (MAPCAs). This combination of PA/VSD/MAPCAs is sometimes referred to as "complex pulmonary atresia" and has been difficult to classify owing to the wide variation in pulmonary blood supply in this rare condition. The condition is part of the family of conotruncal anomalies (which includes TOF, truncus arteriosus, and transposition), and as many as half the cases are associated with CATCH 22 syndrome (cardiac defects, abnormal facial features, thymic hypoplasia, cleft palate, hypocalcemia, and 22q11 microdeletion) [2].

The recent classification of the Congenital Heart Surgery Nomenclature and Database Project [1] (similar to the one of Barbero-Marcial and Jatene [3] in 1990) distinguishes three types of PA/VSD according to the pulmonary circulation: type A with present native pulmonary arteries (NPAs) and no MAPCAs; type B with present NPAs and MAPCAs; and type C with MAPCAs only. Although that goes some way to recognize the heterogeneity of pulmonary blood flow in this condition, there is a need for a more detailed and specific classification system that can be correlated to the management strategies available.

MAPCAs are most commonly associated with PA/VSD but can also be found in other congenital heart diseases [4]. The first mention is attributed to Macartney and colleagues [5] in 1973. An angiographic study published by the Melbourne team found similarities with bronchial arteries, but the nature and origin of MAPCAs is still debated [6, 7]. Most MAPCAs have a thoracic origin (descending aorta and subclavian arteries), but these collaterals can also more rarely arise from cervical vessels [8], the abdominal aorta, and even from the coronary arteries [9]. The number, size, distribution, and course of

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Abbreviations and Acronyms

MAPCA = major aortopulmonary collateral artery
NPA = native pulmonary artery
PA = pulmonary artery
PA/VSD = pulmonary atresia with ventricular septal defect
pLV = left ventricle pressure
pPA = pulmonary artery pressure
pRV = right ventricle pressure
RV = right ventricle
TOF = tetralogy of Fallot

the MAPCAs—as well as their relationship with any NPA system—is at the core of the heterogeneity of the condition. This article seeks to clarify and classify this spectrum.

The successful repair of "simple" PA/VSD was first reported by Lillehei and colleagues [10] in 1955, but the concept of the surgical MAPCAs management did not develop until 3 decades later. Even by 1995, only 25 % of PA/VSD/MAPCAs patients survived to adulthood [11]. However, thanks to the pioneering hemodynamic and histopathologic studies of Macartney, Thiene, Haworth, and their colleagues [12–20] in the 1970s and 1980s, surgical strategies and management have radically changed such that the majority of patients now survive into adulthood with good functional class.

Different philosophies currently exist regarding the best management of this complex and heterogenous group of patients; these will be defined and discussed in this article.

After more than 30 years of surgical experience, an updated review is needed on this difficult and nonconsensual topic. Part of the difficulty comes from the small size of most cohorts and from a confusing terminology. The main objective of this article is to focus on the current surgical management strategies of PA/VSD/MAPCAs (ie, PA/VSD types B and C) and their outcomes. Of note, heart-lung transplantation has been suggested [21, 22] as an option, but this technique will not be discussed in this review as it has been marginally reported and it is not recommended in a routine practice.

Material and Methods

A literature search was performed in December 2017 in PubMed and the Cochrane Library for "pulmonary atresia ventricular septal defect," "aortopulmonary collateral," "pulmonary atresia aortopulmonary collateral," and "tetralogy Fallot aortopulmonary collateral." Only publications in English related to PA/VSD/MAPCAs were analyzed.

Table 1 lists the main studies on the surgical management of patients with PA/VSD/MAPCAs published in the recent era, namely, since year 2000, and constitutes the core of this review. The year 2000 also refers to the year of publication of a landmark article by the Stanford group [23]. This report was an update from another work published 5 years earlier [24] with a cohort of 10 patients that advocated for a midline one-stage repair and unifocalization of PA/VSD/MAPCAs for the first time.

Publications were included in Table 1 according to the following criteria: most recent study by a same institution for a same strategy; consecutive (nonselected) patients with PA/VSD/MAPCAs; overall number of patients in a cohort 30 or more; cohort treated at a single institution; and management toward a biventricular repair.

An exception was made for the article of Bauser-Heaton and colleagues [25]: the study included patients with TOF/pulmonary stenosis and 36% of their patients had previous surgical procedures performed in other institutions. However, the percentage of TOF/pulmonary stenosis was minimal (7.4%), and the management of patients after referral from other institutions was standardized. This exception was also justified by the remarkable size of the cohort (n = 458). Of note, the studied population did not overlap the one from the study of 2000 [23] by the same group (1992 to 1998 versus 2001 to 2016).

Data were extracted when available or calculable (palliation rates).

Results

The largest experiences published for this condition come from Stanford, Birmingham (UK), Melbourne, and Toronto. It has been usual to classify the surgical treatment of PA/VSD/MAPCAs in two dominant and overlapping strategies—unifocalization and rehabilitation—according to the individual pattern and management of MAPCAs. The two concepts are not mutually exclusive, and most researchers recognize a "combined strategy," as introduced by the Melbourne group (Iyer and Mee [26]), that combines the unifocalization and rehabilitation pathways (Fig 1). Most teams have a preferred strategy, although a few groups adopted a mixed management of their patients [27–30].

The Stanford and Birmingham approach has been to incorporate all significant MAPCAs into a policy of early and complete unifocalization—although always utilizing as much of the NPA system as possible. The Melbourne approach has been influenced by the review of the entire historic cohort (n = 82) [31] that found disappointing evolution of unifocalized MAPCAs after a median follow-up of 14.2 years (the longest reported). The consequence was a radical change of the local strategy toward the rehabilitation pathway, focusing on the NPA and avoiding the incorporation of MAPCAs where possible.

The main outcomes are reported in Table 1, which includes 12 studies and their related publications when present. The PA/VSD/MAPCAs were considered repaired when the VSD was closed. One of the key findings from these large series has been the recognition of the heterogeneity in pulmonary blood supply: most patients have a mixed blood supply from a NPA (intrapericardial) system—which is very variable in size and extent—and from a variable number of MAPCAs. The degree to which these two systems overlap is also very

variable, with some areas of the lung having a "dual supply" (ie, fed by both MAPCAs and NPAs) whereas others are supplied exclusively by MAPCAs or exclusively by NPAs. There is a group of 15% to 20% of patients who have complete absence of any NPA [23, 32].

Unifocalization

Unifocalization aims at promoting the growth of MAPCAs without rehabilitating the NPAs. The concept was introduced in 1981 by Haworth and colleagues [18], who suggested to make the "multifocal blood supply unifocal." Unifocalization of MAPCAs was successfully performed only later in the 1980s. The technique consists in anastomosing ipsilateral MAPCAs to each other directly, through patches or through a conduit, and incorporating the ipsilateral native or central pulmonary artery if present (Fig 2A). In a staged procedure, MAPCAs can be unifocalized to a systemic to pulmonary artery shunt (SPS). The procedure can be performed through (postero) lateral thoracotomy or a midline sternotomy. Specific respiratory complications of unifocalization, including airway ischemia [33] and pulmonary reperfusion injury [34], have been reported.

ONE-STAGE UNIFOCALIZATION. The Stanford group (Hanley and colleagues) pioneered the midline unifocalization with repair in a single stage. The first patients underwent this pathway in 1992 [24], after successful case reports by other researchers [35, 36]. In their last publication with 458 patients (the largest cohort to our knowledge), Bauser-Heaton and colleagues [25] reported a repair rate of 88% at a median age of 8.6 months when this strategy was preferentially used for patients. This study showed the highest repair rate and the lowest age at repair of all other experiences combined. Early and late mortality rates were 3.5% and 8.9% respectively. Median right ventricle to left ventricle pressure ratio (pRV/pLV) was 0.40, the lowest reported, expressed as pulmonary artery to aorta pressure ratio (pPA/pAo). However, the median time of measurement of this ratio after repair was uncertain. Estimated survival was 85% at 5 years.

The Birmingham (UK) team [37] have also promoted the one-stage repair with unifocalization as their preferred strategy, using the midline sternotomy approach where possible—but also advocating combined thoracotomy with sternotomy where necessary to access the MAPCAs. Other groups use unifocalization alongside with another strategy: rehabilitation strategy [27] or combined strategy [28, 29].

STAGED UNIFOCALIZATION. The staged unifocalization advocates separate procedures to achieve ipsilateral unifocalization or to recruit vessels in stages to eventually achieve complete unifocalization. That may be helpful when specific vessels are difficult to access through a single approach or if the quality of vessels is poor and a degree of rehabilitation and growth is also required to achieve repair, as will be discussed. An example of this approach is described by Ishibashi and colleagues [38] obtaining an 80% repair rate at a mean age of 8.1 years. Early and late mortality rates were 7% and 15.9%, respectively. This strategy of always using a staged approach is now less popular and no longer followed by many teams [39, 40].

Rehabilitation

Rehabilitation was historically the first strategy applied in PA/VSD/MAPCAs (ie, from the mid 1970s) as an overarching first operation [41–43]. The goal is to promote the growth of hypoplastic NPAs by surgically (or interventionally) generating antegrade blood flow, without intervention on MAPCAs, before reassessing for definitive repair. Rome and colleagues [44] demonstrated in 1993 the feasibility and safety of a rehabilitation strategy only in PA/VSD with hypoplastic pulmonary arteries (excluding patients with absent pulmonary arteries). The concept recognizes the key fact that the NPAs are often hypoplastic in this condition, yet have potentially excellent distribution into the lung parenchyma. Generating good antegrade flow into these native vessels can be uniquely effective in developing growth of the vessels and opening up their distribution.

The recent experience at the Royal Children's Hospital Melbourne [45] showed a repair rate of 82% when the cohort mostly (89%) followed this strategy. There was no early death and 3 (10%) late deaths among 33 patients. After a median follow-up of 4.5 years, the VSD was closed in 73% of patients at a median age of 1.7 years (after a mean of 2 procedures) when patients exclusively underwent the rehabilitation pathway. The median pRV/pLV ratio was 0.64 at a median of 22 months after repair. The concept of rehabilitation has equally been applied to individual MAPCAs where the distribution of the vessel is good but has been deprived of flow (owing to origin stenosis or tortuosity of the vessel, which is often a progressive process). Mobilization of such vessels with or without patch enlargement and then connection to the aorta with a systemic shunt can also be effective in promoting growth. This procedure can be combined with ipsilateral unifocalization, so that the concepts of rehabilitation and unifocalization overlap.

The rehabilitation strategy can be divided into subgroups, according to the first palliation procedure: systemic to pulmonary artery shunt, insertion of the main pulmonary artery in the aorta, or right ventricle to pulmonary artery (RV-PA) connection.

SYSTEMIC TO PULMONARY ARTERY SHUNT. Central shunt (Fig 2B) using a Gore-Tex vascular graft (W. L. Gore & Assoc, Flagstaff, AZ) is the preferred choice in Melbourne, as described by Gates and colleagues [46]. The insertion of a modified Blalock-Taussig shunt represents another option.

INSERTION OF MAIN PA IN AORTA. Iyer and Mee [26] and others [28, 29, 31, 47, 48] originally described an end-to-side anastomosis of the main pulmonary artery to the ascending aorta, the so-called Melbourne shunt (Fig 2C). This type of shunt and strategy was used in reports of selected patients with present native and diminutive central pulmonary arteries, in other words, PA/VSD type

Table 1. Overview of Main Studies on Pulmonary Atresia With Ventricular Septal Defect and Major Aortopulmonary Collateral Arteries Since 2000

First Author [Reference], Location	Year of Publication	Related Previous Publication	Years of Study	Number of Patients	22q11 Deletion Prevalence (%)	Absent CPA (%)	Age at First Surgery (range)	Strategy Preferred	Strategy Subgroup Preferred	One-Stage Repair (% [n/N])
1. Reddy [23], Stanford, CA	2000	Reddy, 1995 [24]	1992–1998	85	14[a]	16	7 m (10 d–37 y)	Unifocalization	One-stage repair	65.9 (56/85)
2. Gupta [48], Los Angeles, CA	2003	Permut, 1994 [66]	1983–2000	104	…	21	1 w (3 d–22 y)	Combined	Melbourne shunt	N/A
3. d'Udekem [31], Melbourne, AU	2005	Iyer, 1991 [26]	1979–1995	82	…	…	1.4 y (7 d–34 y)	Combined	Melbourne shunt	N/A
4. Ishibashi [38], Tokyo, Japan	2007	Sawatari, 1989 [63]	1982–2006	113	26	15	6.3 y (1.1 m–33.8 y)	Unifocalization	Staged repair	N/A
5. Song [39], Seoul, Korea	2009	N/A	1988–2006	40	…	42	8.5 m (6 d–16 y)	Unifocalization	Staged repair	N/A
6. Davies [37], Birmingham, UK	2009	Griselli, 2004 [32]	1989–2008	216	…	…	…	Unifocalization	One-stage repair	51.4 (111/216)
7. Zhu [28], Toronto, Canada	2016	Honjo, 2009 [67]	2000–2013	30	…	16	…	Unifocalization	One-stage repair	93.3 (28/30)
				17			…	Combined	Melbourne shunt	N/A
8. Bauser-Heaton [25], Stanford, CA	2017	Reddy, 1995 [24]	2001–2016	458	36[a]	…	4 m (1.3–7.3)	Unifocalization	One-stage repair	63.9 (186/291)
9. Soquet [45], Melbourne, AU	2017	Liava'a, 2012 [68]	2003–2014	33	39	6	3.3 w (0.4–31.9)	Rehabilitation	Central shunt	N/A
10. Babliak [29], Kyiv, Ukraine	2017	N/A	2007–2009	28	53[a]	18	10 m (2–336)	Unifocalization	One-stage repair	71.4 (20/28)
			2010–2014	55	42[a]	18	9.7 m (1–180)	Combined	Melbourne shunt	N/A
11. Jia [30], Guangzhou, China	2017	N/A	2007–2015	28	…	29	25 m (3–288)	Combined	RV-PA connection	N/A
				24		0	49 m (1–274)	Rehabilitation	RV-PA connection	N/A
12. Trezzi [59], Roma, Italy	2017	Carotti, 1998 [69]	1996–2015	95	38	24	1 y (21 d–14.5 y)	Unifocalization	One-stage repair	67.4 (64/95)

[a] Part of the cohort was missing for calculation of the reported rate. [b] Repair rate, ventricular septal defect closed. [c] One-stage repair. [d] Pulmonary artery pressure to aortic pressure (pPA/pAo). [e] After repair.

CPA = central pulmonary arteries; d = days; ellipses = missing information; m = months; N/A = not applicable; pLV = left ventricular pressure; pRV = right ventricular pressure; RV-PA = right ventricle to pulmonary artery; TNPAI = total neopulmonary arterial index; w = weeks; y = years.

REVIEW

B [49, 50]. The technique has been particularly successful in smaller infants in rehabilitating the NPA system—diminutive-sized central vessels may be more suited to a direct anastomosis as a prosthetic graft can distort or angulate them if they are only 2 mm to 3 mm in size. There is little risk of overcirculation as the vessels are so small that they provide a natural limit on the flow.

RV-PA CONNECTION. The main advantage of this technique is that it sustains diastolic pressure and also delivers predominantly desaturated blood to the lungs (although the VSD is left open, the streaming effect is such that predominantly systemic venous blood will be directed through the conduit) and so achieves more efficient oxygenation than with a systemic shunt. The technique also gives antegrade access to the pulmonary artery tree to perform subsequent interventions such as balloon pulmonary angioplasty. This technique has been promoted by the Birmingham approach [51], and several teams favor this technique as the initial procedure for a rehabilitation strategy [30, 52, 53]. The RV-PA connection can be established in various ways: a transvalvar autologous pericardial patch, a Gore-Tex vascular graft [45, 54], or a homograft or other valved conduits [53]. A valved conduit will reduce the volume load on the ventricle and the use of stiff conduits of fixed size such as the Hancock Dacron conduit (Medtronic, Minneapolis, MN) will maintain predicted flow while preventing the risk of dilation under pressure.

Combined Strategy

Iyer and Mee [26] in Melbourne described a staged approach in 1991, consisting of the promotion

Table 1. (Continued).

Index Used for Decision	Repair Rate[b] (%)	Age at Repair	Early Mortality Rate (%)	Late Mortality Rate (%)	pRV/pLV After Repair	Time of pRV/pLV Measurement	Palliation Rate (%)	Follow-Up	Survival Overall (%)
None	95	7 m (10 d–37 y)	10.6	8.2	0.44[c]	Intraoperative	…	22 m (1 m–69 m)	74 at 4 y
None	56	5.2 y (1–34)	11.5	5	0.55	…	…	10.2 y	83.5 at 10 y
None	65	4 y (3 m–35 y)	8	…	0.62	1.4 years	…	14.2 y (3 m–25 y)	51 at 12 y[e]
None	80	8.1 y (8.3 m–26.7 y)	7	15.9	0.7	Intraoperative	5.3	8.8 y (0.8 m–23.3 y)	69.9 at 20 y
None	42	3 y (4 m–15 y)	3.8	12.5	0.57	Intraoperative	…	54.5 m (13.4 m–205.1 m)	70.5 at 15 y
None	73	…	6	6	…	…	…	2.3 y (0.1 y–6.7 y)	89 at 3 y
None	77	…	4	14	…	…	…	2.2 y (0.02 y–14.1 y)	78.5 at 5 y
	65	…							
None	88	8.6 m (4.9–29)	3.5	8.9	0.40[d]	…	1.9	3 y (0.6 y–7.9 y)	85 at 5 y
None	73	1.7 y (0.2–6.7)	0	10	0.64	22.2 months	10	4.5 y (0.04 y - 8.3 y)	…
Nakata	82	…	10.7	0	…	…	6	7.9 y	92.6 at 9 y
	60	…	0	5.4				3.8 y	
TNPAI	43	…	11.5	13.5	…	…	…		
	62	…						1,145 d (0 d–3451 d)	78 at 5 y
None	67	…	8.4	5.2	0.49	Intraoperative	…	8.5 y (0.01 y–19.9 y)	78 at 15 y

REVIEW

(rehabilitation) of the NPAs, followed by the unifocalization of NPAs and MAPCAs before repair. Gupta and colleagues [48] reported the largest experience with this strategy (104 consecutive patients) in 2003. As discussed previously, many of the reported series that focus on both unifocalization and on rehabilitation will overlap in this combined approach—often needing two or more procedures before repair can be attempted or achieved.

Overlap Between Strategies

There is likely an overlap between the strategies described that may be obvious only to the experienced reader. It is more than likely that patients with small but not extremely diminutive central pulmonary arteries that are connected to most lung segments would have a preparatory procedure to make them grow in all teams. A team operating under a flagship of unifocalization strategy would still categorize this patient as a success of a unifocalization strategy even though the unifocalization involved both MAPCAs and rehabilitated NPAs at the time of repair. Conversely, a patient who underwent a central shunt soon after birth in a team with a flagship of rehabilitation procedure would be classified as a success of this strategy even though an isolated lobar branch perfused by a single MAPCA was connected to the native pulmonary vessels at the time of repair. This patient would be considered a success of unifocalization by the first team.

Concept of Leaving VSD Open

In cases where the quality or distribution of the pulmonary vasculature is poor, then it may not be possible to

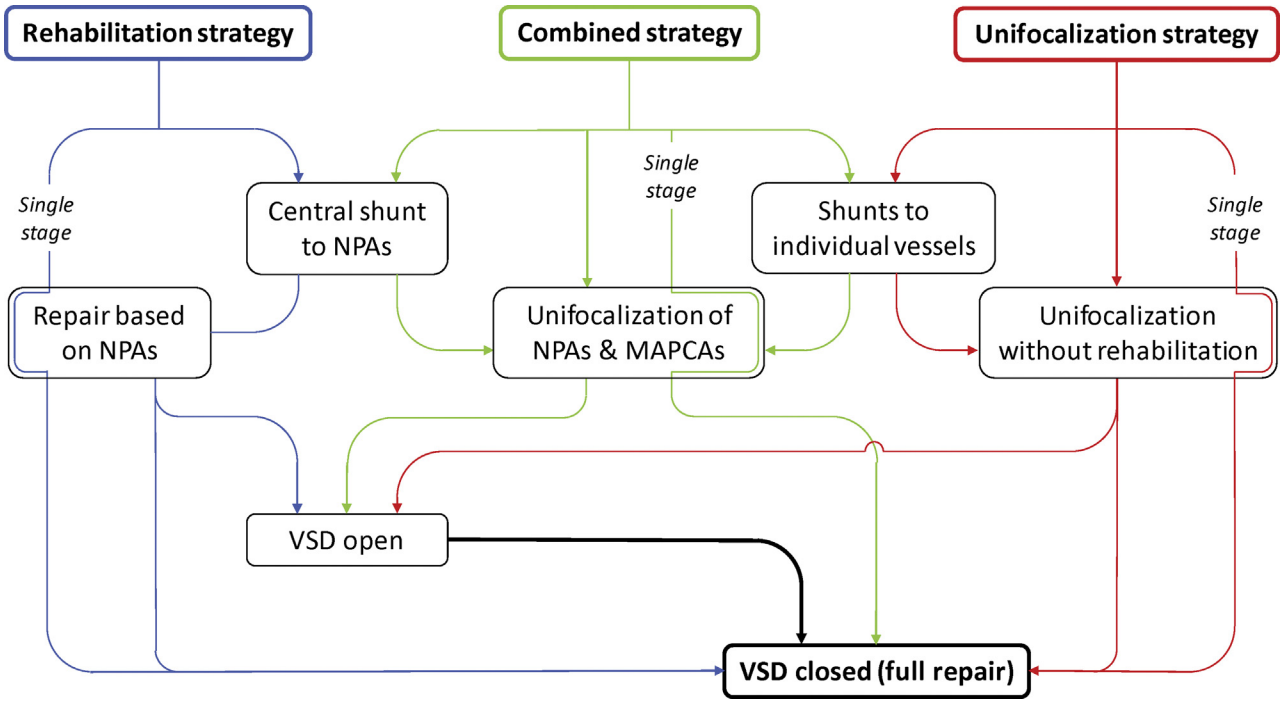

Fig 1. Overview of management strategies for pulmonary atresia with ventricular septal defect (VSD) and major aortopulmonary collateral arteries (MAPCAs). Note: one arrow represents one stage; single-stage repair pathways are represented by a single arrow going through a procedure box before reaching the "VSD closed (full repair)" box. (NPA = native pulmonary artery.)

perform a complete repair (ie, close the VSD and septate the circulation) for risk of suprasystemic RV pressure and RV failure. These decisions are based on the amount of vasculature recruited (minimum 15 of 20 lung segments) or on the intraoperative flow studies. In these situations, there are two options: either the unifocalized vessels are fed by a central shunt, or they can be connected to the heart using an RV-PA conduit that is deliberately restrictive in size. The latter aims to create a situation analogous to a "balanced tetralogy of Fallot" where the restrictive conduit prevents any risk of overcirculation. That has the advantage of sending desaturated blood to the lungs and providing good access for subsequent catheterization—but at the expense of a ventriculotomy. Both approaches have been used successfully, and there is no evidence to suggest one approach is better than the other.

Associated Procedures of PA Enlargement and Interventional Catheterization

All strategies share the necessity of reintervention on pulmonary arteries or MAPCAs by surgery or interventional catheterization procedures. Often hypoplastic before any intervention, pulmonary arteries can show stenosis or hypoplasia in their evolution and may require surgical enlargement with patches or percutaneous dilation and stenting [45]. MAPCAs are also known to stenose or thrombose [15, 16, 31, 36, 39] after unifocalization and can be treated with percutaneous dilation or repeated surgery. In addition, some larger MAPCAs connected to the native pulmonary arteries and causing overcirculation by a dual supply may need to be surgically ligated or coil embolized once antegrade flow has been established [37].

Summary

PATIENTS' CHARACTERISTICS. Studies published since 2000 on PA/VSD/MAPCAs in consecutive series of patients (Table 1) showed a wide range of ages at the time of first surgery (from 3.3 weeks to 6.3 years), depending on the period of treatment and referrals. Of note, the rate of 22q11 microdeletion was reported in approximately half of all cases. The prevalence in recent surgical series was usually 35% to 40%. It has been demonstrated that the anatomy of the pulmonary and systemic-pulmonary vasculature is influenced by 22q11 microdeletion [55, 56] in PA/VSD, generally carrying a negative prognostic value on outcomes [57, 58]. The proportion of patients with absent central pulmonary arteries (PA/VSD type C) varied mostly between 15% and 20%, and rarely exceeded 30% when reported.

STRATEGIES. Unifocalization is currently the most common strategy but the superiority of one strategy over another would be hard to assess at the time of this review. Several teams with extensive published experience have advocated for the one-stage repair [25, 37, 59]. Although earlier referral and earlier unifocalization are associated with the best outcomes, there remains a group of patients at the worst end of the spectrum (10% to 15% of the entire PA/VSD/MAPCAs population) who present with profound cyanosis and poorly developed pulmonary vasculature.

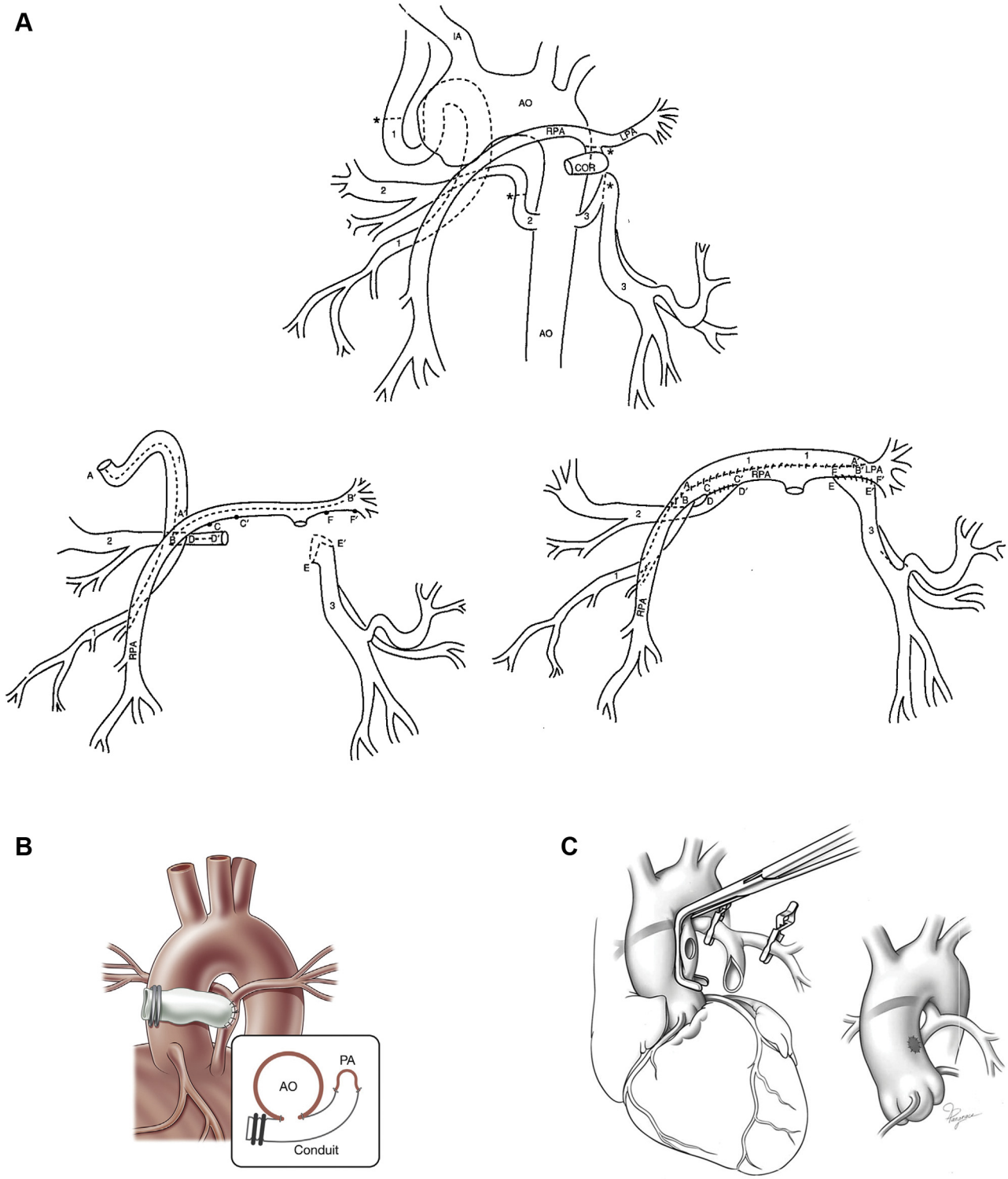

Fig 2. Examples of unifocalization and rehabilitation procedures. (A) Unifocalization of bilateral major aortopulmonary collateral arteries (MAPCAs) directly to the native pulmonary arteries. (Ao = aorta; COR = coronary collateral; IA = innominate artery; LPA = left pulmonary artery; RPA = right pulmonary artery.) (Reproduced from Reddy and colleagues [24] with permission from The American Association for Thoracic Surgery.) (B) Central shunt with a Gore-Tex vascular graft. (Ao = aorta; PA = pulmonary artery.) (Reproduced from Soquet and colleagues [45] with permission from The Society of Thoracic Surgeons.) (C) Central aorta to pulmonary shunt (Melbourne shunt). (Reproduced from Mumtaz and colleagues [49] with permission from The Society of Thoracic Surgeons.)

Single-stage unifocalization can rarely be achieved in these patients, and palliative shunts, ipsilateral unifocalizations, or Melbourne shunt are often all that can be offered initially for this group.

The Stanford group performed repair in 88% of patients at 8.6 months, with a very low early mortality rate (3.5%) [25]. The Birmingham approach reported 2.8% early mortality among 249 patients undergoing unifocalization and repair [51], with a further 22 patients (8.5%) remaining palliated with shunts. The Melbourne study achieved repair in 82% of patients with no operative mortality at an age of 1.7 years, which is early considering that this strategy is staged [45]. With a combined staged strategy involving 60 patients, Babliak and colleagues [29] had a repair rate of 60% with no early mortality.

There is no doubt that NPAs provide the best platform on which to base ultimate repair and that patients who have recruitable (or rehabilitatable) NPAs will have the best long-term results. Nevertheless, there are cases where there is complete absence of NPAs—yet unifocalization and repair has been achieved using MAPCAs exclusively. Furthermore, many cases will have a mixed pulmonary blood supply coming from both NPAs and from MAPCAs, and the results from Stanford and Birmingham would suggest that unifocalization of both components yields excellent results.

Different indexes have been suggested to estimate the size and cross-sectional area of the pulmonary or neopulmonary arborization [60–62]. Although those indexes have been repeatedly reported [28–30,38,39], the recent publications listed in Table 1 interestingly reveal that they have been very rarely involved in the decision-making process [29, 30]. Most groups decided on the progression of their patients according to a suggestive estimation of the size of the pulmonary tree measured by the percentage of segments of the lung perfused (images of angiography or computed tomography) [37, 45] or according to an intraoperative flow study [23, 25, 28, 59].

OUTCOMES. Depending on institutions and strategies, the repair rate varies between 42% and 95%. The age at repair was missing in several of the reported studies. Early mortality for patients with PA/VSD/MAPCAs was acceptable, with a rate of less than 11.5% all institutions, strategies, and procedures combined, and inhospital mortality of less than 5% in the larger series. After repair, pRV/pLV was not always measured, and apart from intraoperative measurements, the time between repair and measurement of the ratio was specified in only two studies (by the Melbourne group). The overall survival rates were mostly between 78% and 85% at a range varying from 3 years to 20 years after the first surgery.

In all series, there is a small number of patients who still cannot be repaired owing to the poor development of NPAs or unifocalized MAPCAs, regardless of the strategy. This category of patients is unlikely to undergo a septation and is left palliated, cyanotic but alive. As seen in Table 1, these data were rarely mentioned. When not available but clearly stated, the proportion of palliated patients was calculated according to the method described in Figure 3 [25, 29] and varied from 1.6% to 10%. The category of patients "awaiting repair" is not included in the calculation process because of the uncertain fate (repair, death, or palliation), even when repair is likely.

Comment

This review offers an updated comparison of the main studies on PA/VSD/MAPCAs published since 2000. Of note, all publications were retrospective. We have demonstrated that the surgical management of PA/VSD/MAPCAs has evolved into three main strategies: unifocalization, rehabilitation, and a combined strategy. This review also shows a great heterogeneity in patients treated and deficiencies in data reporting.

Unifocalization, Rehabilitation, and Combined Strategy

The strategy described by Iyer and Mee was named "staged repair" by the original researchers and others [47, 48], and was classified as being a part of the unifocalization group. After having described the unifocalization and rehabilitation strategies earlier in this review, it seems obvious that the term "staged repair" can be confusing for any reader because it can also refer to a subgroup of the unifocalization strategy [38, 63] as well as to rehabilitation because of the inevitable staged nature of the latter.

Common Themes

The Stanford team highlighted that some neonates require early repair when presenting in heart failure [64]. Heart failure, especially early in life, implies well-developed pulmonary vasculature and suitability for early biventricular repair. In many instances, these patients have a limited number of collateral arteries, and single-stage repair with unifocalization is feasible with excellent results. All teams seem to have accepted this strategy.

Also, when small central pulmonary arteries are present and seem to be distributed to most of both lungs, most teams will grow these pulmonary arteries by either a central shunt, a Melbourne shunt, or restoration of the RV-PA continuity. Although it is not clear from publications by teams who have adopted unifocalization strategies, because the details of the operative procedures are often unreported, this practice is currently common.

A technical aspect that has become clear is that the unifocalized vessels should be dissected from the aorta and mobilized as distally as possible because the collateral segment of these vessels can considerably remodel and become stenotic. This dissection is by far more easily done in a virgin field, and all efforts should be made for that dissection to be performed only once. MAPCAs are frequently located very posteriorly in the mediastinum, and division of their origin with extensive mobilization can help bring the vessels further forward and avoid

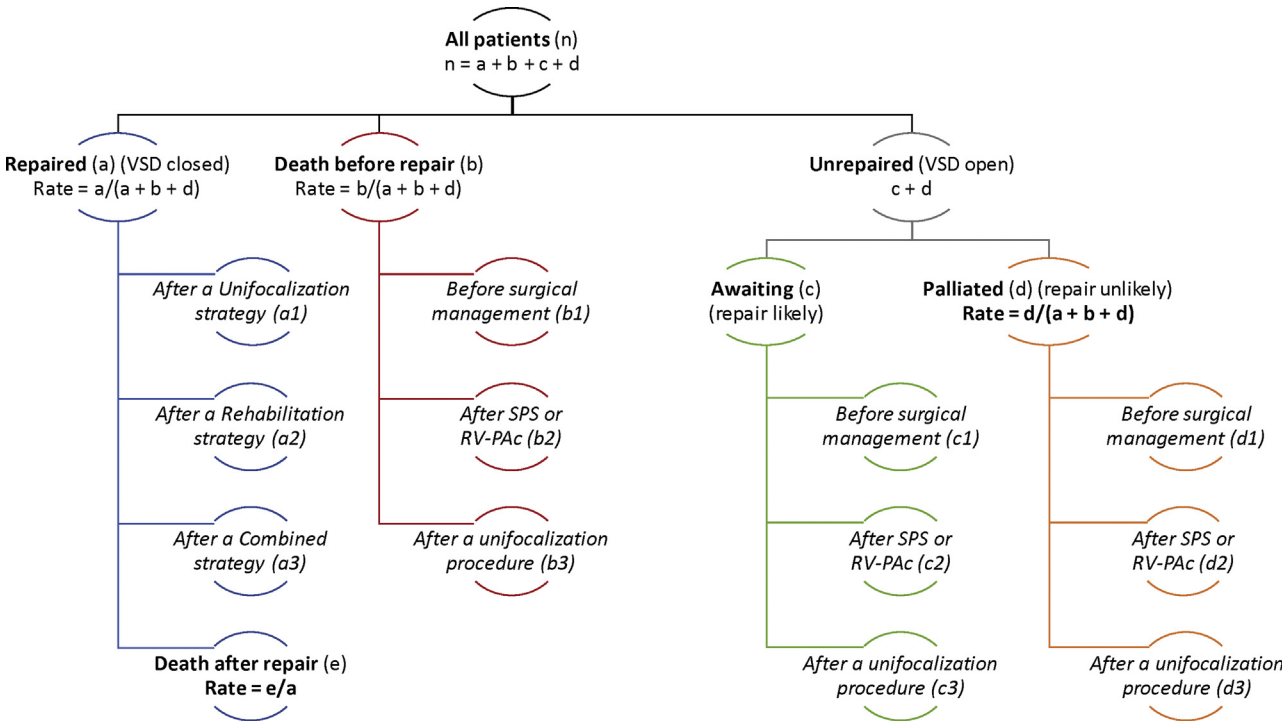

Fig 3. Recommended presentation of outcomes regardless of strategy. (RV-PAc = right ventricle to pulmonary artery connection; SPS = systemic to pulmonary shunt; VSD = ventricular septal defect.)

compression on the neighboring airways. Also, identification of the vessels within the fissures of the lung can provide better access, requiring confident and extensive mobilization of the lungs that is not a feature of most cardiac surgeries and which demands considerable experience and confidence with the anatomy. Detailed three-dimensional imaging with computed tomography and magnetic resonance imaging is essential to define the relationship of the vessels with the main airways and esophagus.

INVESTIGATIONS. An extensive imaging study is necessary at presentation of all patients. Injections of each MAPCA in angiographic studies are essential to define pulmonary blood supply and to establish areas supplied solely by MAPCAs and those with dual supply. Pulmonary vein wedge injections may reveal "orphaned" vessels that had lost their initial blood supply but fill retrogradely and can be recruited. As an alternative, computed tomography scan or magnetic resonance imaging can be performed before surgery and have the advantage over angiographic studies to be able to define the relationship of the vessels to the airways and esophagus.

PALLIATION. Whether some patients are better left palliated at least in the first years of life remains an open question. Patients at the most extreme end of this spectrum of the disease may be those at higher risk of early mortality, such as infants with absent or diminutive central pulmonary arteries and multiple collaterals with no connection to any intrapulmonary vessel and distal near-normal arborization. These patients may survive many years with a good quality of life. Some investigators report that late unifocalization of some of these patients after several years of palliation can be quite successful, alleviating the challenges of performing this surgery in smaller infants [31].

WHEN UNIFOCALIZING, SHOULD WE UNIFOCALIZE EVERYTHING?. Most teams will proceed with unifocalization in a varying proportion of patients. Even those prone to follow a strategy of rehabilitation will unifocalize neonates in heart failure and patients failing the rehabilitation strategy. When faced with diminutive collateral vessels, the question arises as to whether they are best left alone or unifocalized. An area of debate remains as to whether a staged approach to such vessels may be better or whether there is a critical size or area of distribution at which recruitment has no long-term benefit.

Long-Term Outcomes

Aiming to achieve septation wherever possible is a laudable approach but risks leaving some patients with a hypertensive RV and at risk of developing RV dysfunction and failure. Patients with pulmonary vasculature at the poorer end of the spectrum may be better managed by leaving the VSD open and utilizing shunts or limiting RV-PA conduits as long-term palliation. This approach may give very good long-term quality of life in selected patients. Finding the

threshold at which each strategy meets the best long-term outcome for these patients is a further question to be answered through better classification of the spectrum of disease.

Moreover, as survival has nowadays become acceptable, we would be able to compare the various strategies, not only in terms of combined morbidity including number of surgeries and repeated operations or interventional catheterization procedures, but also in terms of functional capacity and quality of life.

Recommendations for Authors

An important observation extracted from this review is the variability in the reporting of data. Without greater standardization of reporting results, it will be difficult to ascertain the superiority of techniques and approaches [65]. Given our analyses, we recommend that all the following data, listed in Table 1, should be reported: prevalence of 22q11 microdeletion, proportion of patients with absent central pulmonary arteries (PA/VSD type C), pre-repair procedures and timing, age at repair, post-repair pRV/pLV ratio and pPA/pLV ratio and their time of measurement, and proportion of patients left palliated indefinitely, calculated according to the described method (Fig 3).

Attention should also be paid to the anatomic characterization of central pulmonary arteries (presence, confluence, size) and MAPCAs (presence and number, size, origin, course and supplied pulmonary segments, connection with the native pulmonary arteries—dual supply).

Most reports today are alluding to the general philosophy of repair followed by each center, but insufficient data are provided to identify the best strategy and procedures in the various subset of patients composing this complex condition.

We propose that every patient be given an individualized code that represents their exact anatomy and a second code that describes the procedures (see Table 2 for definition of MAPCAs and proposed standard format for recording current patient status): (1) MAPCA coding: for each lung, the number of MAPCAs with notation as to whether they are sole supply or dual supply; size is indicated by number of segments supplied. (2) Native PA coding: complete absence as a separate code; if present, then size and distribution to each lung (size: diminutive, small, moderate). (3) Procedure: one-stage complete repair. (4) One-stage unifocalization (to shunt or to RV-PA conduit). (5) Ipsilateral unifocalization or shunt procedure to MAPCA/PA only.

Staged approaches would then list the subsequent procedures with a similar code: second shunt, complete repair, unifocalization only, and so forth. Therefore, a patient could be described as having three MAPCAs: one with dual supply to left lung; one with sole supply to left lung (three segments); and one with dual supply to right lung. The NPAs were present but small to the left and moderate to the right. This patient might have undergone procedure 1, unifocalization with RV-PA conduit (VSD left open) of native PAs plus recruitment of one MAPCA and ligation of two MAPCAs; or procedure 2, complete repair.

Conclusion

The surgical management of PA/VSD/MAPCAs has evolved into three primary surgical strategies. Outcomes are excellent with both the rehabilitation strategy and the unifocalization strategy (more than 80% achieving full septation). There remains some debate over the value of always recruiting all MAPCAs, but the best results are achieved when MAPCAS are recruited in the absence of dual supply or poorly developed native pulmonary arteries. Investigators should be encouraged to

Table 2. Proposed Definition of MAPCAs and Standard Format for Recording Current Patient Status

MAPCA Definition	Example
Each vessel coded by	MAPCA #1, right subclavian artery, RUL, three segments, sole supply
Origin ± bifurcation[a]	MAPCA #2
Position	#2a, descending aorta, RML, two segments, dual supply (with RPA)
Number of segments supplied	#2b, descending aorta, LUL, two segments, sole supply
Sole supply or part of dually supplied segment	MAPCA #3, and so forth
Standard format for recording current patient status	
Shunt procedures to individual vessels	
Can be subclassified into those with subsequent unifocalization anticipated and those seen as final destination	
Shunt or RV-PA connection to native PAs	
Can be subclassified as those with subsequent unifocalization anticipated and those seen as final destination	
Unifocalized but ventricular septal defect left open	
Can be further subclassified by whether subsequent full repair is anticipated	
Can also be subclassified by whether unifocalization was achieved in single stage or after previous shunt	
Unifocalized and ventricular septal defect closed (ie, full repair)	

[a] One major aortopulmonary collateral artery (MAPCA) that bifurcates would be coded #Xa and #Xb.

LUL = left upper lobe; PA = pulmonary artery; RML = right middle lobe; RPA = right pulmonary artery; RUL = right upper lobe; RV = right ventricle.

REVIEW

standardize the reporting of their data to better elucidate optimal strategies and procedures.

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