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Outcomes of Complete Repair of Mixed-Type Total Anomalous Pulmonary Venous Return

Mohannad Dawary[1], Fareed Khouqeer[1], Ziad Issa[1], Louai Alkhalaf[2], Faisal Alshamdin[3], Massimo Griselli[2]

[1]King Faisal Specialist Hospital and Research Center, Clinic of Cardiac Surgery, Riyadh, Saudi Arabia

[2]Princess Nourah bint Abdulrahman University, King Abdullah Bin Abdulaziz University Hospital, Clinic of Cardiac Surgery, Riyadh, Saudi Arabia

[3]AlJouf Cardiac Center, Prince Mohammed Bin Abdulaziz Medical City, Clinic of Cardiac Surgery, Riyadh, Saudi Arabia

Abstract

Objectives: To retrospectively review 14 cases, describe preoperative imaging, anatomic findings and confirmation at surgery, surgical technique, and outcomes.

Materials and Methods: We describe anatomic variations in mixed-type total anomalous pulmonary venous return and surgical outcomes in a case series from 2 centers. Mixed-type total anomalous pulmonary venous return is classified based on the pattern of pulmonary venous drainage. Type I refers to '2+2' drainage of venous pairs, type II refers to '3+1' and type III refers to all other variants. Six patients (43%) had type I "2+2" drainage; 2 patients had 2 supra-cardiac and 2 cardiac connections, and 4 patients had 2 infra-cardiac connections and 2 cardiac. 4 patients (29%) had type II "3+1" drainage. Three patients had 3 cardiac and 1 supra-cardiac variants and one had a rare supra-cardiac pulmonary venous combination. 4 patients (29%) had type III morphology. 2 patients of them had 3+2 anatomy with 3 supra-cardiac and 2 cardiac connections, both of which included 3 right-sided pulmonary veins. The remaining 2 patients had unique anatomy, one with tri-level attachment to cardiac, supra-cardiac, and infra-cardiac and the last with all supracardiac pulmonary venous drainage but in a "3+1+1" pattern.

Address for Correspondence: Massimo Griselli, Princess Nourah bint Abdulrahman University, King Abdullah Bin Abdulaziz University Hospital, Clinic of Cardiac Surgery, Riyadh, Saudi Arabia

e-mail: mgriselli63@gmail.com **ORCID:** orcid.org/0009-0003-9520-7944

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Abstract

Results: In previous publications, mixed-type total anomalous pulmonary venous return has a higher mortality rate, with univariate analysis of mixed-type versus cardiac type with statistically significant hazard ratios of 2.88 in previous study and mortality as high as 42-50%. We achieved improved outcomes, with overall survival of 79% and no intraoperative mortality. Both patients who had complex intracardiac lesions (1 with ventricular septal defect and 1 with tetralogy of fallot) ultimately died. Of the remaining patients, 29% developed pulmonary venous obstruction on echocardiography follow-up with 3 requiring interventions with cardiac catheterization.

Conclusion: Mixed-type total anomalous pulmonary venous return has a wide variety of connections, which contributes to the complexity of planning and surgical correction. Further studies are needed to better understand the different morphologies of the disease.

Keywords: Mixed type, outcome, surgical management, total anomalous pulmonary venous return

Introduction

Total anomalous pulmonary venous return (TAPVR), also referred in literature as total anomalous pulmonary venous connection (TAPVC), provides a wide spectrum of complex anatomic and therapeutic challenges amongst congenital heart disease. An estimated 400 infants (1/10.000 births) are born annually in the United States with TAPVR, an incidence among congenital heart disease of 0.7-1.5%[1,2]. While there may be rare survival to adulthood if un-repaired[3,4], typical median survival if unrepaired is 2 months with 50% mortality in first 3 months of life, thereby demonstrating the urgency of recognition and intervention[4].

TAPVR is classified according to the Darling system developed in 1957 into cardiac, supracardiac, infracardiac, or mixed-type depending on the pattern of pulmonary venous (PV) drainage to the systemic venous circuitry[5]. Cardiac forms have PV return to the coronary sinus or right atrium (RA) directly. Supracardiac forms have PV return to the superior vena cava (SVC) or innominate veins (often via a vertical or ascending venous connection from the confluence). Infracardiac forms include PV return to various inferior structures including the inferior vena cava, portal venous system or hepatic veins (often via a descending inferior vein which penetrates the diaphragm). Mixed-type TAPVR involves 2 or more of these variances. The latter of these is the most rare type, ranging typically from ~11%-12.3%[6,7] in various single-center studies, though as low as 3-4%[8,9] and as high as 20-21%[10,11]. TAPVR can also be separated based on cardiac defects. Simple (or isolated) TAPVR may include an additional atrial septal defect (ASD) or patent ductus arteriosus, while complex TAPVR includes at least one other complex cardiac defect[12].

Mixed-type TAPVR can be further classified based on pattern of PV drainage. Type I refers to '2+2' drainage of venous pairs, type II refers to '3+1', and type III refers to all other variants[13]. This classification may be significant as the risk of death is 5.8 times higher in type III in one study[14]. Mixed-type TAPVR in itself has a higher mortality, with univariate analysis of mixed type versus cardiac type (lowest mortality of TAPVR variants) demonstrating statistically significant hazard ratios of 2.88 in previous study[7], and mortality as high as 42-50%[15,16]. Other determinants of increased mortality or transplant in this study include complex TAPVC and postoperative length of stay, while older age, weight greater than 2.5 kg, non-emergent operation, lack of PV obstruction, cardiac and supracardiac types were associated with increased survival to discharge[7]. Additionally, prolonged cardiopulmonary bypass (CPB) time and infracardiac involvement have been correlated with increased risk of mortality[15].

Overall TAPVR mortality may be decreasing with time[7], which may be partially due to earlier recognition and a change in preoperative diagnostic strategies over

recent decades. Complex mixed-type TAPVR has now been demonstrated in fetal ultrasound as early as 18 weeks[17]. There has been a move towards more advanced post-natal imaging including the use of angiography. An earlier clinical study of TAPVR cases from 1983-1993 demonstrated confirmatory diagnosis (following 2D echocardiogram with color doppler) by catheterization or magnetic resonance imaging in only 17/36 patients, the other 19 diagnosed intraoperatively or by autopsy[18]. For reference, sensitivity and specificity of echocardiography for anatomic diagnosis in mixed type TAPVR may approach 31% and 100%, versus 94% and 99% in cardiac catheterization[19]. This is not without risk, however, and the value and importance of angiography in preoperative planning due to the complex variance of mixed-type TAPVR anatomy is increasingly recognized[20,14]. A recent study by Turkvatan et al.[12] demonstrated 100% diagnostic agreement between preoperative low-dose multidetector computerized tomography angiography (CTA) and intraoperative findings, including all 5 of their mixed-type patients (supracardiac and cardiac anomalies referenced). Similarly, Fulanetto et al.[10] reference CTA with 3D reconstruction assisting with improved outcomes (8% mortality in their mixed-type cohort which included 3 type III variants).

While TAPVR literature has continued to compound, there remains relatively limited mixed-type TAPVR-specific case series. The following is a retrospective clinical review. We aim to describe and display the anatomic variance and surgical technique used in the operative repair of one surgical group's experience with 14 mixed-type TAPVR patients between 2004-2018 in an attempt to add to the comprehensive available data with which clinicians can use to optimize decision-making in a rare congenital lesion bearing significant mortality.

Materials and Methods

The Institutional Review Board approved the data collection for this study (approval number: 2245682, date:18.12.2024) and the need for patient consent for enrollment and publication was waived because of the retrospective design. Literature review via PubMed search was performed with combinations of the following keywords: 'TAPVR', 'TAPVC, 'mixed type', and 'surgical technique'. Patient data was reviewed to outline objective data listed in tables IV-VII and artistic visualizations were provided to aid with interpretations and overall understanding of lesions. Illustrations were drawn by professional audio-visual expert.

Statistical Analysis

As our study is a retrospective, we used descriptive statistics for all the continuous variables were reported as mean ± standard deviation while categorical variables were reported as frequencies and percentages.

Surgical Technique

All patients received satisfactory endotracheal anesthesia and sterile prep. Visualization was made in all cases via a standard median sternotomy followed by opening of pericardium in midline and dissection for confirmation of TAPVR anatomy. Patients were heparinized, cannulated, and placed on complete blood profile (CBP). Aorta cross-clamped and cooling targeted mild hypothermia or deep hypothermia cardiac arrest depending on anticipated case difficulty. Dual caval snares were placed except if procedure required avoidance of PV distribution on SVC. Right atriotomy was typically made for cardiac subtypes. Ascending veins if present were either doubly ligated or ligated near confluence and reflected back to left atrium (LA), details which are further described in morphologies section. Autologous pericardium was used when needed for redirecting venous drainage, wall closure, or septation, with only two exceptions where bovine pericardium was used (patients L and N). Details of repair were overall dependent on variation in mixed type anatomy and outlined elsewhere in the morphology section. Suture type most frequently used included 5-0, 6-0, and 7-0 prolene or polyprolene, though also included PDS and Maxon variants. Venae cava snares were then released. Rewarming performed

with warm blood cardioplegia while venting through the aortic root. Pressure monitoring lines were inserted in few cases where hemodynamics deemed necessary. Weaned off CPB once normothermic, then decannulated once stable. Protamine given for heparin reversal then chest tubes and temporary pacer wires placed once hemostasis obtained. Sternum was either closed in layers or left open depending on complexity of repair. If left open, gap in skin was covered with an Esmarch patch and sewn to skin edges with 4-0 nylon. Wounds were cleaned and dressed appropriately. Instrument, sponge and needle counts were carried out twice.

Complications

There were few procedural complications. Patient F had bleeding due to incisions to upper and lower L PVs, felt likely made during dissection. This was corrected via autologous pericardium for upper vein and primary repair for lower vein. Two other procedures required extended duration due to findings at conclusion of their case, both of which had significant other cardiac comorbidities. Patient L had continued narrowing at SVC repair site which was subsequently augmented with a patch of bovine pericardium, however after weaning off CBP it was felt to still be unoptimized so required a 2nd pump run for additional bovine patch repair. Finally, patient H had tension at arch anastomosis site noted during rewarming, requiring deep hypothermia circulatory arrest and arch reconstruction. This was followed by TEE noting SVC obstruction at RA junction, for which SVC was re-implanted to right atrial appendage without CBP.

Morphologies

(Type I): 2+2

Six patients (43%) displayed type I morphology (Table 1). All of these included supracardiac connection, 4 had cardiac connection (pts A, B, F, G), 2 had infracardiac connection (pts D, M). One supracardiac connection was on the right (R pulmonary veins to SVC; pt F), otherwise all supracardiac connections involved L pulmonary veins draining to the innominate vein via a confluence and vertical vein. Cardiac connections varied, 2 of which included R PVs to coronary sinus (pts A, G), 1 with R PVs to SVC/RA junction (pt B), 1 with L PVs via confluence

Table 1.

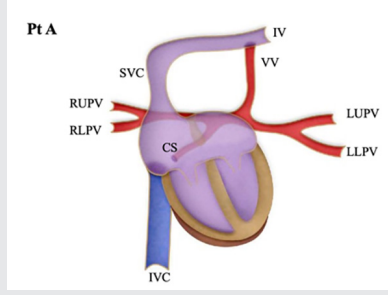
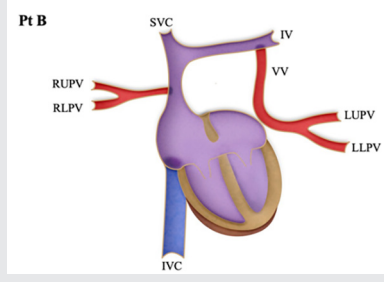
Patient ID	Anatomical variations	Illustration
Pt A	Type I: 2+2 Supracardiac connection + cardiac connection	
Pt B	Type I: 2+2 Supracardiac connection + cardiac connection	

Table 1. Continued

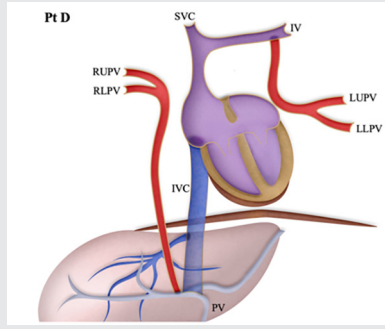
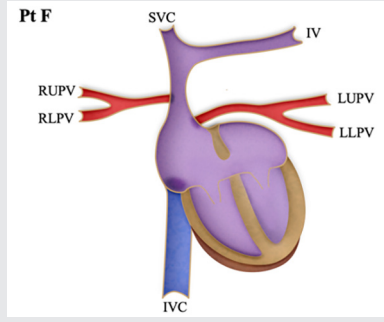
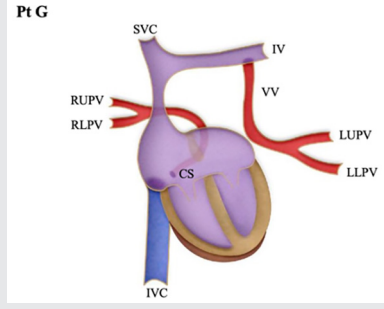
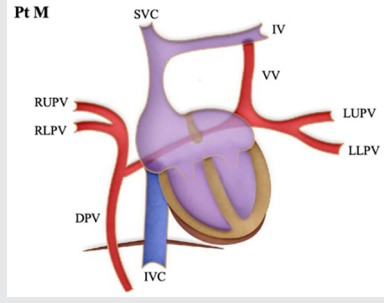
Patient ID	Anatomical variations	Illustration
Pt D	Type I: 2+2 Supracardiac connection + infracardiac connection	
Pt F	Type I: 2+2 Supracardiac connection + cardiac connection	
Pt G	Type I: 2+2 Supracardiac connection + cardiac connection	
Pt M	Type I: 2+2 Supracardiac connection + infracardiac connection	

Illustration of different anatomical variations of type I for patients A, B, D, F, G, and M. RUPV: Right upper pulmonary vein, RLPV: Right lower pulmonary vein, SVC: Superior vena cava, CS: Coronary sinus, IVC: Inferior vena cava, IV: Innominate vein, VV: Vertical vein, LUPV: Left upper pulmonary vein, LLPV: Left lower pulmonary vein, PV: Portal vein, DPV: Descending pulmonary vein

into RA (pt F). One of the coronary sinus variants had a connection between the R and L venous confluences, which was utilized during surgical technique (pt A). Infracardiac connections included a R descending vein connecting R PVs to portal vein (pt F), and one unusual connection with a R confluence which drained below the diaphragm towards the liver, as well as transversely to a L PV confluence (pt D).

Of the 5 L vertical veins, 4 were doubly-ligated and divided, while one was ligated and reflected for anastomosis of L PVs to LA (pt G). For the L PV confluence to RA, the confluence was close enough to the LA that mirrored incisions were made for direct anastomosis at that location (distal to confluence ligated and removed). For the two coronary sinus attachments, the sinus was unroofed for drainage into LA. Of these, the patient with a connection between their R and L confluences had their L vertical vein doubly ligated and removed, leaving all pulmonary veins to drain through the coronary sinus (confirmed unobstructed). The SVC-RA connection was directed to the LA via autologous pericardium. One of the infracardiac variants had the left and right confluences connected to each other after bilateral vertical vein ligation and detachment, following which both were attached together to the LA (pt D). The latter infracardiac variant mentioned (pt M) had each PV confluence attached to the LA separately (each via 6-0 PDS suture), otherwise similarly having ligation and division of other vertical veins as well as the inter-confluence connection.

(Type II): 3+1

Four patients (29%) displayed type II morphology (Table 2), all of which were cardiac/supracardiac variants (pts I, K, H) with the exception of a rare supracardiac/supracardiac combination (pt C). The left upper pulmonary vein (LUPV) emptied into the innominate via a vertical vein in all but one case (pt H) where it emptied with the L and R LPVs into the RA [in this pt the right upper pulmonary vein (RUPV) emptied into the SVC]. Two of the other PV trios drained into the coronary sinus (pts I, K), while one combination drained into the SVC (this being the supracardiac/supracardiac variant).

Table 2.

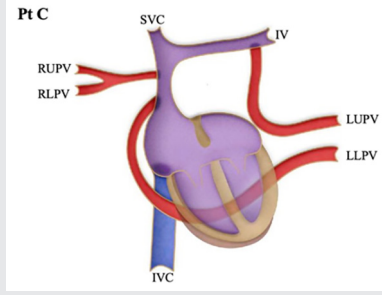
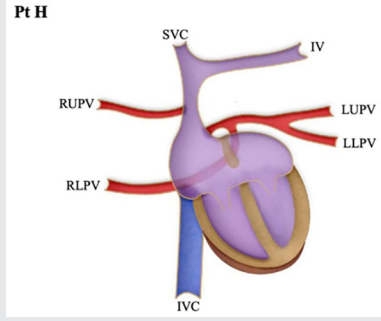
Patient ID	Anatomical variations	Illustration
Pt C	Type II: 3+1 supracardiac/supracardiac combination	
Pt H	Type II: 3+1 cardiac/supracardiac variants	

Table 2. Continued

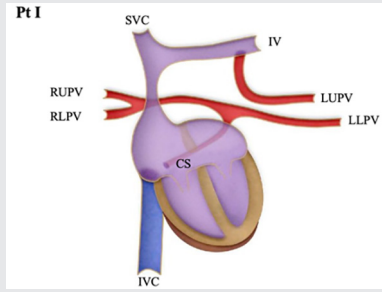
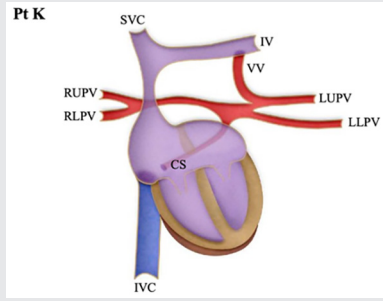
Patient ID	Anatomical variations	Illustration
Pt I	Type II: 3+1 cardiac/supracardiac variants	
Pt K	Type II: 3+1 cardiac/supracardiac variants	

Illustration of different anatomical variations of type II for patients D, I, J, L. RUPV: Right upper pulmonary vein, RLPV: Right lower pulmonary vein, SVC: Superior vena cava, CS: Coronary sinus, IVC: Inferior vena cava, IV: Innominate vein, VV: Vertical vein, LUPV: Left upper pulmonary vein, LLPV: Left lower pulmonary vein

Details regarding the repair of the supracardiac-supracardiac morphology were unavailable (pt 'C'). Vertical veins when present for LUPV to innominate connection were singly ligated and reflected back to the LA (including pt who had an additional but obstructed connection from LUPV to confluence). Coronary sinus was unroofed followed by autologous pericardial ASD closure in both cases with confluence drainage into sinus. In the patient with PV drainage to the SVC and RA, the atrial septum was fully excised to an existing sinus venosus ASD followed by autologous pericardial patch redirection of all PVs into the LA. As noted in the complication section above, this repair did result in SVC-RA obstruction ultimately requiring re-implantation of SVC to R atrial appendage (pt H).

(Type III):

Four patients (29%) displayed type III morphology (Table 3). Two of these patients had 3+2 anatomy with supracardiac and cardiac connection, both of which included 3 right-sided pulmonary veins (pts N,L). The first of these had 2 small RUPVs which emptied into the SVC, while the LPVs and the right lower pulmonary vein (RLPV) emptied directly into the RA (pt N). The second (pt L) - who also had tetralogy of fallot (TOF) - had LPVs drain via vertical V to a retroaortic innominate V, with more complex variance in the R sided veins (RLPV to LA, RMPV and RUPV to SVC). The remaining two patients had rather unique anatomy. One of these (pt E) had a mixed variant including a tri-level attachment: cardiac, supracardiac (which was inferred, as true drainage was unclear), and infracardiac connection. The RPVS attached to a multitude of locations, including the RA, a PV confluence, and a descending infra-diaphragmatic vein. The LVs attached more classically to PV confluence, though also with a vertical vein which was difficult to determine exact location of emptying. The last patient (J) had LPVs and RLPV all attached to the SVC-RA junction via a L confluence, while the RMPV directed into the RA and RUPV into the SVC (resemblance of a '3+1+1').

Surgical repair was dependent on lesion, however technique largely involved similar strategies to those used in types I and II. Decision was made not to intervene on the tiny RUPVs on the first of the above 3+2 variants (pt N), while the remaining RA attachments were re-routed to the LA by a bovine patch following ASD enlargement. The other 3+2 variant had a complete ligation of vertical vein and anastomosis to LA, while the SVC was opened for disconnection of the RUPV and RMPV, which were re-anastomosed to the LA. Autologous pericardium was

Table 3.

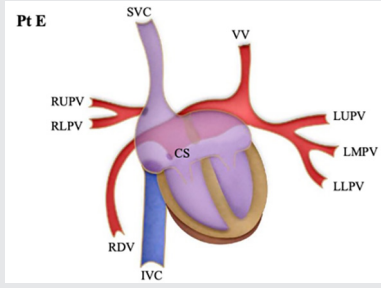
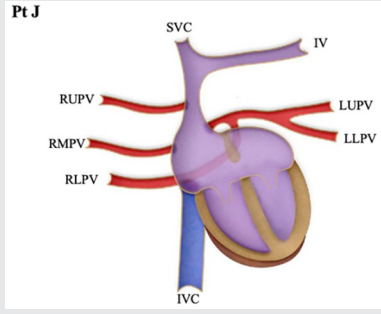
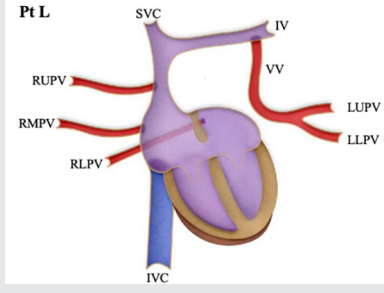
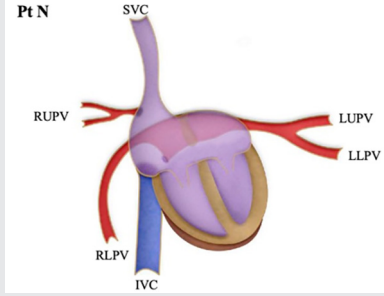
Patient ID	Anatomical variations	Illustration
Pt E	Type III: Mixed variant including a tri-level attachment: cardiac, supracardiac and infracardiac connection	
Pt J	Type III: LPVs and RLPV all attached to the SVC-RA junction via a L confluence, while the RMPV directed into the RA and RUPV into the SVC	
Pt L	Type III: 3+2 anatomy with supracardiac and cardiac connection	
Pt N	Type III: 3+2 anatomy with supracardiac and cardiac connection	

Illustration of different anatomical variations of type III for patients F, K, M, O. RUPV: Right upper pulmonary vein, RLPV: Right lower pulmonary vein, RDV: Right descending vein, RMPV: Right middle pulmonary vein, SVC: Superior vena cava, CS: Coronary sinus, IVC: Inferior vena cava, IV: Innominate vein, VV: Vertical vein, LUPV: Left upper pulmonary vein, LLPV: Left lower pulmonary vein, LMPV: left middle pulmonary vein

used to direct SVC flow to RA. This was all in addition to a complete TOF repair. Similarly, the '3+1+1' variant had their SVC extensively opened (to the innominate V) to accommodate a large autologous pericardium for redirection of the RUPV and RMPV to the LA. The confluence containing the LPVs and the RLPV was otherwise detached and anastomosed to the LA. The pt with tri-level attachment had classic incision and anastomosis of the L confluence to the LA. The remaining RA drainage was redirected into the same LA anastomosis via elongation of the atrial incision, followed by ASD closure.

Findings

Baseline demographic data is outlined in (Table 4). Of note, all patients were under 6 moths old except for two (10 moths, 132 moths), exaggerating the mean age and weight some. All three patients who died had weights greater than 4 kg at initial operation.

Pertinent preoperative data are outlined in (Table 5). Regarding mortality, 1/7 patients who underwent catheterization, 1 of the patients who received computerized tomography angiogram, and the patient who had 3-D printing performed ultimately died. Despite this, 3-D printing was deemed by the surgical team to be helpful in facilitating discussion regarding surgical approach. Patients with complex intracardiac lesions both died as well. Interestingly, none of the three patients who had obstruction indicated in operative note deceased, a number that would be discrepant from much of the literature. Pt 'N', whose course is outlined further below, developed obstruction later and did die. Even in these 4 patients, 25% obstruction mortality may still be low in relation to available data in some mixed type TAPVR

Table 4. Baseline data

	% Male (n)	% Female (n)	
Sex	50 (7)	50 (7)	
	Median	Mean	Range
Age (mo)	3.2	12.4	0.26-132
Weight (kg)	4.335	5.775	2.7-27.7
BSA (m2)	0.25	0.275	0.17- 0.7

Table depicting baseline demographic value for all 14 patients in series. mo: Month; kg: Kilogram; m2: Meters squared, BSA: Body surface area

Table 5. Preoperative data

Workup performed	% present (n)	Detail
Echocardiogram	100 (14)	
Cardiac catheterization	50 (7)	
CTA	14 (2)	Pts: K, N
Cardiac MRI	7 (1)	Pt: M
3-D print reconstruction	7 (1)	Pt N
Anatomic findings	% present (n)	Detail
Obstruction	21 (3)	Pts: B, C, D, N
Complex intracardiac lesions	14 (2)	Pt H = VSD Pt L = TOF
Non-complex intracardiac lesions	71 (10)	ASD in 10 of these, PDA in 9
Other anomalies	29 (4)	PHTN, b/l SVC, Ao arch hypoplasia, pleural-pericardial communication

Table depicting preoperative values for all 14 patients in series. CTA: Computed tomography angiogram, MRI: Magnetic resonance imaging, Pt: Patient, VSD: Ventricular septal defect, TOF: Tetralogy of fallot, ASD: Atrial septal defect, PDA: Patent ductus arteriosus, PHTN: Pulmonary hypertension, b/l: Bilateral, Ao: Aorta

cohorts, which can approach 100%[19]. As is discussed in limitations, the sample size of this series is not able to conclude any association with these findings.

Intraoperative data are presented in (Table 6). Circulatory arrest was reported in 6 patients, one of which was discarded (reported as 1-minute, unclear validity). Prime volume was also only occasionally reported. Temperature ranges for mild, moderate, and deep hypothermia were determined based on Saad citation [21], though others site different ranges. Both of the patients whose bypass times were greater than 4 hours ultimately died (cross-clamp times greater than 2.5 hours). There did not appear to be any recognizable patterns with temperature regulation, drainage, or monitoring devices.

Postoperative data is presented in (Table 7). There appeared to be no pattern regarding initial admission length, including pt 'J' whose stay was ~6 months and remains alive as of this report. It should be noted that pt 'N' had a rather extensive and complex course that included re-operation due to baffle dehiscence of original ASD, and 2 months later for PV obstruction from cor triatriatum membrane. Patient was also the only readmission (failure to thrive) which was complicated by pulmonary edema with profound respiratory failure felt to

Table 6. Intraoperative data

	Median	Mean	Range
CPBT	118 min	136 min	65- 270 min
XCT	69.5 min	85.7 min	36-176 min
Circulatory arrest Time (n=5)	43 min	38.8 min	20- 51 min
Prime volume (n=5)	650 mL	607 mL	500-700 mL
Temperature (n=13)	% present (n)	Detail	
Mild hypothermia	31 (4)	32°, 34°	
Moderate hypothermia	15 (2)	28°	
Deep hypothermia	54 (7)	16°, 18°	
Devices placed (n=13)	% present (n)	Detail	
Mediastinal CT	92 (12)		
Pericardial CT	46 (6)		
Pleural CT	85 (11)	One unilateral (Pt: I), all others b/l	
Peritoneal catheter	46 (6)		
LA pressure line	15 (2)	Both also had pulmonary artery pressure monitor	
RA pacer wires	100 (13)		
RV pacer wires	23 (3)		

Table depicting intraoperative values for available patient data. CPBT: Cardiopulmonary bypass time, XCT: Cross-clamp time, min: minute, mL: Milliliter, mild hypothermia: 32-35°C; moderate hypothermia: 26-31°C, deep hypothermia: < 26°C; CT: Chest tube, b/l: bilateral, LA: Left atrium, RA: Right atrium, RV: Right ventricle

Table 7. Postoperative data

	Median	Mean	Range
Initial hospital stay (n=13)	13 d	45.4 d	9-193 d
	% present (n)	Detail	
Mortality	21 (3)	Pts: C, L, N Time to death: 1 mo, 2 mo, 6 mo	
Post-op echo obstruction	29 (4)	1 spontaneous resolution on follow-up echo; 3 w/o obstruction mentioned pre-op	

Table depicting postoperative values for available patient data. d: Days; mo: Month, Echo: Echocardiogram

be contributed to by PV obstruction. Of note, this patient also had one anomalous PV which was not manipulated at initial repair, as mentioned above. Two-staged repairs have been successfully reported in the literature, though have involved avoidance of LUPV repair[22,23]. It is unclear whether this contributed at all to this patient's demise. Postoperative echocardiograms revealed obstruction in 4 patients who did not have it preoperatively, for unclear reasons other than in pt 'N'. Two of these patients ('N', 'H') are deceased. Anatomically, one of the deceased patients was type II ('H'), the others type III ('L', 'N'). There was no mortality in type I connections.

Discussion

This study outlines the complex variance in mixed type TAPVR patients, and to our knowledge only 5 other mixed type-specific retrospective case series [15,24,10,14,25] exist in the literature, though we acknowledge mixed type data can be found within generalized TAPVR studies. The intent of this data was to add to the literature for purposes of future meta and systematic analyses to improve surgical approach, as well as serve educational purposes regarding a rare and intricate lesion.

Interestingly, in this cohort there was no intraoperative mortality. This appears low in comparison to data in available studies, including a 768-patient cohort by Shi et al.[26] which included 38 intraoperative deaths (5%), though this article did not include mixed-type specific data. Other studies site operative mortality rates for mixed-type TAPVC of 19.3%[14] and up to 50%[16], though the latter included 2/5 deaths which included single ventricle anatomy as a comorbidity. Our cohort did not include any single ventricle anatomy, however one patient did have complex congenital disease (tetralogy of fallot) and ultimately did not survive. Additionally, both patients who had complex intracardiac lesions (per mixed type TAPVR classification) died (tetralogy of fallot, ventricular septal defect). This is one of three total mortalities (rate of 21%), all occurring within the first year. This rate is lower than that of a similar cohort, citing 42% in first year[15]. It is difficult to account for this difference, as our cohort included a higher percentage of both infradiaphragmatic connections as well as type 3 lesions, both of which are deemed to have higher mortality rates. This is also a lower rate than a larger TAPVR analysis, where mixed type mortality rate was 30%[7].

Aside from the complex intracardiac lesion finding related to mortality, the following were other pertinent observations regarding the available data as outlined above. Patients who died were all greater than 4 kg at operation (5 patients were under 4 kg). No patients with preoperative obstruction at initial surgery died. 3-D print reconstruction was performed in 1 patient and deemed helpful for initial repair, though ultimately patient did die. Both patients with bypass times greater than 4 hours (cross-clamp times greater than 2.5 hours) died. The other, as previously mentioned, had a short bypass time, but prolonged and complex course including two additional operations (baffle dehiscence, cor triatriatum repair).

Study Limitations

There were notable limitations in this study. Given that this was an observational retrospective cohort, there is no correlational data. The majority of our data collected (aside from outcomes) from our cohort is from operative notes, relying on accuracy of the note writer and consistency of full completion. Certain data was also unavailable to be retrieved when analysis performed, specifically patient C who's operative note was not located. Also, one of the patients operated on was at a center in a different country than the previous 13 (patient N), lending potential for institutional differences to contribute to outcome data. Lastly, given small sample size the results are difficult to make meaningful inferences from, however this is consistent with previous cohorts given the rarity of this disease, and contribution to a future systematic review may prove effective.

Conclusion

Mixed type TAPVR presents a wide variety of connections, lending to the complexity in planning and

execution of surgical correction. Our study contributes further to the available data on this more rare form of TAPVR, which may be important as more cohorts emerge with varying success rates. More widespread studies, including systematic review may be beneficial to further characterize morphology, intraoperative technique, outcome data, and perhaps most importantly, specific indices for predicting higher survival or mortality rates.

Ethics

Ethics Committee Approval: The Institutional Review Board approved the data collection for this study (approval number: 2245682, date:18.12.2024).

Informed Consent: The need for patient consent for enrollment and publication was waived because of the retrospective design.

Footnotes

Authorship Contributions

Surgical and Medical Practices: Dawary M, Khouqeer F, Issa Z, Alkhalaf L, Alshamdin F, Griselli M, Concept: Dawary M, Khouqeer F, Issa Z, Griselli M, Design: Dawary M, Khouqeer F, Griselli M, Data Collection and/or Processing: Dawary M, Issa Z, Alkhalaf L, Alshamdin F, Griselli M, Analysis and/or Interpretation: Dawary M, Khouqeer F, Issa Z, Alkhalaf L, Alshamdin F, Griselli M, Writing: Dawary M, Khouqeer F, Griselli M.

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