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Stent interventions guided by three-dimensional rotational angiography to treat total cavopulmonary connection stenosis☆

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ABSTRACT

Background: Stenosis of the total cavopulmonary connection (TCPC), in particular the pulmonary arteries, is common in children with single ventricle physiology. Stenting is an efficient interventional strategy to treat stenosis and optimize TCPC hemodynamics.

Methods: A retrospective single center study was performed to investigate the prevalence, causes and outcome of TCPC stenosis in children treated with stent implantation guided by three-dimensional rotational angiography (3DRA).

Results: From September 2011 to February 15th 2021 48 patients received 73 stents at 67 locations in 59 3DRA stent procedures. Median age and weight were 3.6 years (0.0–17.2) and 14.1 kg (3.4–70.0), respectively. Left pulmonary artery (LPA) stenosis accounted for 75% of the stenosis and is often caused by retroaortic compression. Adverse events occurred in 6 catheterizations (10.2%). Twenty-one patients (43.8%) underwent 27 reinterventions including planned serial redilation (N = 17), extra stent placement (N = 6), surgery (N = 2), unplanned balloon dilation (N = 1) and stent placement (N = 1) for restenosis by intima proliferation.

Conclusion: TCPC stenoses, especially of the LPA, are common in children with single ventricle physiology and external compression is one of the main causes. 3DRA safely guides stent interventions as intraprocedural knowledge of possible interactions with surrounding structures are evaluated. Besides it is effective in both short and long term and can be performed in small children. Restenosis by intima proliferation is rare, though repeated redilations are necessary to match stent diameter with patient size.

1. Introduction

Stenosis of the total cavopulmonary connection (TCPC), in particular the pulmonary arteries (PA), is common in each stage of single ventricle palliation [1–3]. Unobstructed flow through the extracardiac tunnel, caval veins and pulmonary arteries (PA) is necessary for optimal TCPC physiology, peripheral PA development and long-term clinical outcome [3–6]. Key factors in PA stenosis are primary hypoplasia, anastomotic scarring from shunt or patch plasty [4], size of the Norwood aortic arch patch plasty [6], size and position of the original PA bifurcation [7] and external compression by the close interaction of the aorta, pulmonary artery and airway trias [8]. Possible risk factors for extra cardiac conduit stenosis are type of material, diameter and length [9,10].

Early detection, understanding and treatment of TCPC stenosis is essential for optimal cardiac preload and to minimalize congestive complications and eventually TCPC failure [2]. Surgical treatment is invasive and might cause or exacerbate vessel stenosis [4]. The close interaction of the aortic-pulmonary-airway trias often makes the PAs inaccessible or unsuitable for surgery, making a percutaneous approach the treatment of choice [3].

Stenting has shown to be effective for tunnel stenosis [10]. In addition, it is more effective to release PA stenosis (up to 100%) than balloon dilation (60%, recurrence rate up to 35%) by reducing recoil and effects of external compression [3,4,6]. However, reinterventions are necessary in case of intima proliferation or to match stent diameter with patient growth to an adult size [3,4,6,11]. In our center, three-dimensional rotational angiography (3DRA) has become the standard of care for the evaluation and treatment of TCPC stenosis in the majority of patients.

This study investigates the prevalence, causes and outcome of TCPC stenosis in children treated with 3DRA guided stenting in our center.

☆ The authors take responsibility for all aspects of the reliability and freedom from bias of the data presented and their discussed interpretation.

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Abbreviations

3DRA = three-dimensional rotational angiography
DKS = Damus-Kaye-Stansel
LPA = left pulmonary artery
PA = pulmonary arteries
PCPC = partial cavopulmonary connection
PLE = protein losing enteropathy
RPA = right pulmonary artery
SVC = superior vena cava
TCPC = total cavopulmonary connection

2. Methods

2.1. Study design

A retrospective study was performed at our institution in children with single ventricle physiology stage I to III that underwent stent procedures of their TCPC with use of 3DRA since the introduction of this technique in September 2011. Stent procedures of the caval veins, extracardiac tunnel, pulmonary arteries and innominate vein were included. Stent interventions of shunts, the arterial duct or the aorta were excluded. Procedural consent was given by the parents or patient. Our local institutional review committee approved this study and waived individual consent for data collection.

2.2. Procedural description

All cardiac catheterizations were performed under general anesthesia by the same team of clinicians. 3DRA imaging was performed on the Artis Zee system with *syngo*DynaCT (Siemens, Forchheim, Germany) and use of our univentricular heart protocol [12]. All patients received complete hemodynamic assessment, including pulmonary wedge pressures and pressure pullback loops. 3D reconstructions were made from the rotational angiographic images and included the inner chest structures and interactions between vessels and surrounding tissues. These 3D reconstructions were back-projected on the frontal plane and used for roadmap guidance of the stent intervention. After stent implantation control angiography and pressure pullback loops were performed to detect vessel trauma and prove unrestricted flow. Echocardiography was performed one day after catheterization before hospital discharge.

2.3. Definitions

Stent interventions within 6 months after surgery were defined as early. Morphological success was achieved if the lumen diameter was normalized (post-stent lumen diameter/pre-stent lumen diameter $\geq$1) [13] and hemodynamic success if there was no residual pressure gradient [14]. Adverse events were defined as major when being life-threatening if left untreated (e.g. cardiac arrest, major bleeding or device embolization) or minor as not being life-threatening, transient and un-anticipated (e.g. transient arrhythmia or balloon rupture) [3,13,15].

2.4. Data collection

Baseline data as well as indication for cardiac catheterization and procedural characteristics were collected. Follow-up data on patient outcome after first 3D guided stent placement, stent patency, re-interventions, cardiac surgery and adverse events were collected. Data are presented as frequencies or medians with range.

3. Results

3.1. Demographics

Between September 2011 and February 15th 2021, 272 3DRA catheterizations were performed in 136 children in stage I to IIII, with 186 interventional procedures (68.4%). Stenosis of the caval veins, pulmonary arteries, innominate vein or extracardiac tunnel were treated with stents in 48 patients during 59 of these procedures (31.7%), in which 73 stents were implanted at 67 locations. Median age and weight at intervention were 3.6 years (0.0–17.2) and 14.1 kg (3.4–70.0), respectively. Left pulmonary artery (LPA) stenosis was the most frequent substrate: 50/67 locations (74.6%) (Table 1). Twenty-two patients underwent previous surgical or percutaneous interventions of their TCPC, with PA patch plasty (N = 16), balloon dilation (N = 14) and stenting (N = 14) (Table 2).

3.2. Indication for catheterization and stent placement

3DRA is routinely performed at our department before partial cavopulmonary connection (PCPC) and TCPC. Other indications were: stenosis (N = 30), plastic bronchitis or protein-losing enteropathy (PLE) (N = 5), acute post-operative hemodynamic failure (N = 4), evaluation of aortic arch (N = 4), fatigue or desaturations during exercise (N = 2), recanalization of innominate vein (N = 3), chylothorax with unknown cause (N = 2) and closure of collaterals (N = 1). In the majority of stenosis (45/67, 67.2%) the pressure gradient related to the morphological substrate. In the other cases stents were placed for either aberrant vessel morphology (N = 19) or on clinical indication (N = 3).

3.3. Causes of stenosis

LPA compression due to an oversized aorta was frequent after Norwood or Damus-Kaye-Stansel (DKS) procedure (N = 23). Aortic arch or coarctation stenting caused LPA compression in two patients. In one case the LPA was surgically positioned anterior to the DKS thus compressed by the neo-aorta from posterior. Surgical clips on a shunt or duct caused LPA stenosis in three and superior caval vein stenosis in 1 out of 43 patients. In one child with right atrial isomerism, severe bilateral pulmonary vein stenosis occurred after bilateral anastomosis with the appendices.

Table 1
Patient demographic and procedural characteristics.

Variable	Value
Number of patients (male)	48 (28)
Number of catheterizations	59
Number of stenting procedures; stents placed	67; 73
Left pulmonary artery	50; 54
Right pulmonary artery	5; 6
Tunnel/Extra cardiac conduit	4; 5
Superior caval vein	5; 5
Innominate vein	3; 3
Age at stent implantation (years)	3.6 (0.0–17.2)
Body weight (kilograms)	14.1 (3.4–70.0)
Stage at intervention	
Pre-PCPC	2
PCPC	23
TCPC	34
Pre-interventional pressure gradient	Gradient (mmHg); remark
Left pulmonary artery (46)	2 (0–5); 7 patients without gradient, 1 complete occluded vessel
Right pulmonary artery (3)	3 (0–4); 1 patient without gradient
Tunnel (4)	1 (0–2); 2 patients without gradient
Superior caval vein (2)	1 (–)
Innominate vein (3)	NA; 2 complete occluded vessels

Data are presented as numbers or medians with range (min-max). PCPC = partial cavopulmonary connection; TCPC = total cavopulmonary connection.

Table 2
Previous interventions.

Type (N)	Location (N)	Stage (N)	Age in years	Time to 3D stent in years
PA Plasty (16)	Both (12), LPA (2), RPA (2)	pre-PCPC (2), PCPC (13), TCPC (1)	0.50 (0.03–2.34)	5.37 (0.11–15.94)
Balloon (14)	LPA (11), RPA (2), IV (1),	pre-PCPC (2), PCPC (7), TCPC (5)	1.55 (0.12–10.56)	2.17 (0.00–9.91)
Stent (14)	LPA (12), SVC (1), Tunnel (1)	pre-PCPC (2), PCPC (10), TCPC (2)	0.74 (0.06–4.99)	5.05 (0.77–10.64)

Data are presented as numbers or medians with range (min-max). IV = innominate vein; LPA = left pulmonary artery; PA = pulmonary artery; PCPC = partial cavopulmonary connection; RPA = right pulmonary artery; SVC = superior vena cava; TCPC = total cavopulmonary connection.

Stenting of both pulmonary venous ostia with stents of 8- and 10-mm diameter was successful but did lead to severe right pulmonary artery (RPA) compression necessitating RPA stenting.

Other causes of stenosis were relative stent stenosis due to growth (N = 4), thrombus formation with total vessel occlusion needing recanalization (innominate vein N = 3, LPA N = 2, extracardiac tunnel N = 1) and in-stent stenosis based on intima proliferation (N = 1).

3.4. Stenting technique

All patients were evaluated with 3DRA prior to stenting, including reconstruction of surrounding structures. In 19 procedures an additional 3DRA with balloon interrogation was necessary to evaluate the risk of airway or coronary artery compression when stenting the target substrate. In one case bronchoscopy was used simultaneously with balloon interrogation to evaluate airway patency. In another case a conventional angiography was performed in addition to 3DRA to assess the distance between LPA and coronary arteries.

The optimal angulation for visualization of each stenosis was analyzed based on the 3DRA reconstruction. A cranial-caudal angulation of more than 45° was necessary in 55% of the cases for best understanding of the stenotic substrate (Fig. 1), which exceeds the limits of the C-arm in monoplane setting.

3DRA guided stenting at stage I was performed in two patients. The first patient was a 10-day old newborn who underwent a Norwood operation two days earlier. Antegrade RPA stenting was performed through a Sano shunt with a Biotronik kinetic energy coronary stent 5 × 15mm delivered through a 5 Fr guide catheter. The other patient was 3,5 weeks old and had a total occluded LPA that had to be recanalized and stented three weeks after Norwood operation. A 2.6 Fr Ninja Swift mini steerable catheter with a 0.014-inch PT guidewire was used to enter the occluded LPA antegrade through the central aorto-pulmonary shunt. Subsequently the LPA was stented a 4.5 × 13 mm Cre8 coronary stent.

Stenting at stage II was divided in early (N = 11) and late procedures (N = 12). Stents were placed early after PCPC at a median of 40 days (5–134) or late before TCPC at 1.2 years (0.8–3.4 years). All stents were placed through jugular vein access with a median sheath size of 6 French (6–8). After TCPC completion four stenting procedures were early performed at a median of 35 days (23–134) after surgery. In the other 30 patients late stent implantation was done at a median of 5.5 years (0.7–14.7) after TCPC. Femoral vein access with a median sheath size of 8 French (5–12) was used for stent placement in TCPC patients with the exception of four PA stents and one IV stent that were placed through the jugular vein with a 7 French (6–9) sheath.

Extracardiac conduits were stented in four patients, including both Vascutek (N = 2) and Goretex (N = 2) tunnels. The innominate vein was completely occluded in two of these patients necessitating recanalization. This was managed with a telescope technique (Teflon tipped coronary guide wire, 2.5 Fr braded microcatheter, 4 Fr multipurpose tapered tip diagnostic catheter and 5Fr guide catheter). After predilatation with coronary balloons a BeGraft peripheral covered stent 8 × 37mm (Bentley, Germany) was implanted followed by ultra-highpressure balloon dilatation and flaring. Details of the stents, balloons and catheters used at stage II and III are listed in Table 3.

All stenting procedures were successful without residual pressure gradients. None of the stents dislocated and stent strut dilation for stent jailing was necessary in 9 cases. In this series no stent fractures were observed. In twenty-six cases the post-interventional result was evaluated with 3DRA.

Three major adverse events occurred in 59 procedures (5.1%), all three caused by a thrombus in the pulmonary artery during LPA stenting, which were immediately and successfully removed by thrombosuction. Although all patients received heparin 100IE/Kg per protocol, two patients had activated clotting times (ACT) < 200 s when thrombus formation was detected. Minor adverse events that occurred were moderate

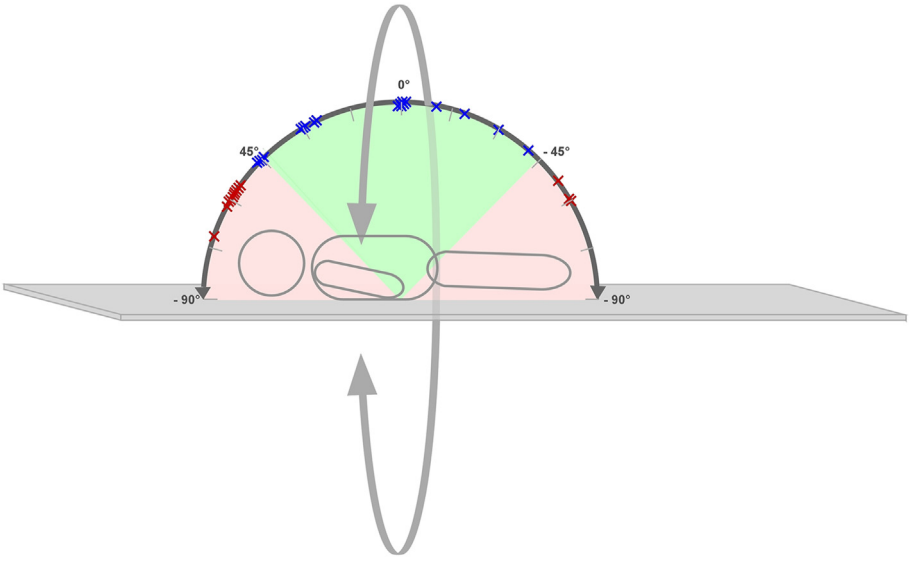

Fig. 1. Cranio-caudal angiographic angulations for diagnosis and intervention of PA stenosis. The optimal cranio-caudal angulations to diagnose PA stenosis exceeded the 45° range of the C-arm (green area) in more than half of the patients (red area/red crosses). The best anterior-posterior angulations for RPA and SVC stenosis was −30° and for tunnel stenosis −45° (grey arrow).

Table 3
Procedural details per location and stage.

Location	N	Length (mm)	Diameter (mm)	Balloon
LPA (PCPC)				
Cook Formula 535	13	16–26	4–8	Advance
EV3 Mega LD	7	16–26	8–10	Powerflex
Cook Formula 418	2	12–20	6–8	Advance
LPA (TCPC)				
Cook Formula 535	7	20	6–10	Advance
EV3 Mega LD	23	26–36	8–18	Powerflex, CBV Balt (N = 5), Advance (N = 4)
EV3 Max LD	1	26	20	CBV Balt
RPA (PCPC)				
Cook Formula 535	1	16	8	Advance
Atrium ADVANTA V12[a]	1	16	8	Powerflex
RPA (TCPC)				
Cook Formula 535	2	12–16	6–8	Advance
EV3 Mega LD	1	26	10	Powerflex
SVC				
Cook Formula 535 (PCPC)	4	12–16	6–8	Advance
EV3 Mega LD (TCPC)	1	26	12	Powerflex
IV (TCPC)				
Bentley[a]	2	37	8–10	(premounted)
Cook Formula 535	1	20	7	Advance
Tunnel				
Andra XXL	2	48–57	15–16	CBV Balt
EV3 Max LD	2	36	16	CBV Balt
CP[a]	1	39	16	Atlas

[a] Covered stents. IV = innominate vein; LPA = left pulmonary artery; PA = pulmonary artery; PCPC = partial cavopulmonary connection; RPA = right pulmonary artery; SVC = superior vena cava; TCPC = total cavopulmonary connection.

bronchial compression after stenting in two patients and a retroperitoneal hematoma in one case.

3.5. Reinterventions

During follow-up 27 patients (56.3%) were free of reintervention after stent placement. In the other 21 patients 27 reinterventions were performed (Fig. 2).

The patient with early RPA stent placement after Norwood operation went for PCPC after 3.5 months with patch plasty of the RPA and partial stent removal. Two days later the patient underwent an emergency bail-out stenting procedure. In the other patient with stenting pre-PCPC, the LPA stent was removed and a PA patch plasty was performed during PCPC surgery. 3DRA guided restenting of the LPA because of lasting stenosis was performed 4,5 months later.

Redilation of stents implanted after PCPC was performed in 10 patients with a median of 1.4 years (0.1–6.1) after stent placement. Stents implanted after TCPC had to be redilated at 3.0 years (0.3–5.8) in 5 patients. Indication for LPA redilation was a morphological substrate with pressure gradient of 1–3 mmHg due to somatic growth (N = 11), intima proliferation (N = 1) or both (N = 3). Somatic growth was the only indication for superior vena cava (SVC) (N = 3) stent redilation. In the patient with right atrial isomerism, bilateral pulmonary vein stents and RPA stent, redilation of all stents was performed because of somatic growth. Current diameters of the pulmonary venous stents are 10 and 12 mm.

The initial stent placed in stage II was elongated with a second one in three patients after a median of 1.1 years (0.9–6.0). In two patients stent elongation was indicated at 2.9 years (1.0–4.7) after the primary stent was implanted at stage III. Seven patients underwent 8 catheterizations with stent placement on other locations in TCPC than the initial stent. Six of these were 3DRA guided and described within the results.

Stents were longitudinally cut open to enable TCPC completion in 11/22 patients (LPA N = 10, RPA N = 1).

3.6. Clinical outcome

At a median follow-up of 4.7 years (0.7–9.5) after the initial 3D guided stent intervention 44 patients were alive and clinically doing well. Echocardiography showed stent patency during routine evaluation in the outpatient clinic in 36/44 patients. In the other patients the retro-aortic LPA stent itself could not be visualized but perfusion of periphery was normal. In the patient with right atrial isomerism the pulmonary vein stents are patent with low pressure gradients under vitamin-K antagonists 9.5 years after initial implantation. Unfortunately this patient developed recurrent PLE shortly after TCPC procedure 6 years ago and is currently stable on PLE medication. Four patients suffered from PLE pre-interventionally which resolved after stent placement and transient medication in two patients, but is still ongoing in the other two patients who are on medication. A relapse of PLE was seen in a patient after balloon dilation, but she is currently free of symptoms. Finally, one patient suffered from plastic bronchitis although receiving long-term low

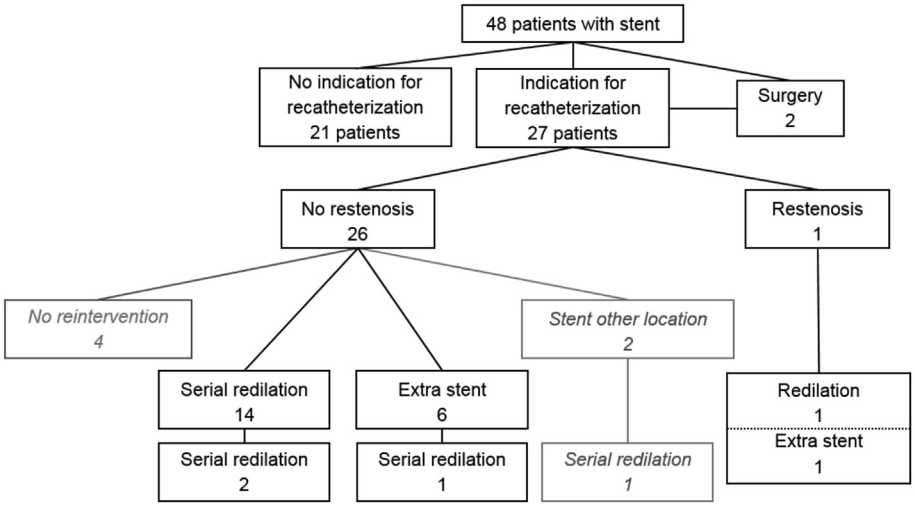

Fig. 2. Recatheterization and reinterventions after initial stent placement. The recatheterization rate was 56.3% (27/48). Two patients with stent placement pre-PCPC underwent both surgery and recatheterization. In one patient intima proliferation caused restenosis necessitating redilation and extra stent placement. In the other 26 patients, four were free of reintervention and two went for stent placement on a different location (boxes in grey, not included in reintervention number). The other 20 patients had reinterventions as serials redilation or extra stent placement, resulting in a reintervention rate of 43.8% (21/48).

molecular heparin therapy. After stenting of a significant LPA stenosis heparin therapy could be ceased without recurrence of casts.

During follow-up four patients died (8.3%). One patient died after Norwood procedure due to an anastomosis rupture during bail-out stenting 2 days after PCPC. Two other patients after PCPC had a fatal outcome despite bail-out stent procedures with massive thrombotic occlusions in one and global hemodynamic instability in another patient. The last patient underwent mechanical aortic valve implantation but did not survive the complex postoperative course due to severe rhythm disorder.

4. Discussion

Single ventricle physiology is characterized by passive pulmonary artery perfusion driven by intrathoracic pressure changes due to respiration. Any obstruction in TCPC affects this passive blood flow and might cause congestive complications in the long term. In our center 3DRA is the standard imaging method to evaluate TCPC and guide interventions, as it accurately visualizes cardiovascular anatomy within its context of surrounding structures. This study evaluates the prevalence, causes and treatment outcomes of TCPC stenosis guided by 3DRA. The results show that in our population 35% of the patients suffer from TCPC stenosis, with LPA stenosis being the most common substrate (75%) caused by external compression of the aorta after Norwood or DKS surgery. Besides, anastomosis stenosis and extracardiac conduit obstructions can also limit the efficiency of the passive pulmonary perfusion. 3DRA is an ideal modality to guide stent procedures in any stage of single ventricle palliation, as interactions with other intrathoracic structures are visualized and taken into account enhancing procedural safety. Clinical outcome is promising in both short and long-term, improving TCPC hemodynamics and securing PA development. In addition, stents can be placed in small children, tackling the problem of vessel stenosis in an early and crucial phase of vessel development. Restenosis by intima proliferation is rare, though serial redilations are necessary to match stent diameter with patient size to an adult size.

4.1. Causes of stenosis and stenting technique

Stenosis of the extracardiac conduit is rare and occurred in 4 patients (out of our population of 129 with extracardiac tunnel; 3.1%). In our institution Vascutek prothesis were abandoned a decade ago due to severe intima proliferation with 67% failure [9] and those were replaced by Goretex conduits in most patients to create the extracardiac tunnel. Stent implantation with 3DRA guidance was performed in the only two patients left with Vascutek prosthesis. Goretex conduits were stented in one case with a twisted conduit and another case with Goretex anastomosis stenosis.

Severe obstruction of the superior caval vein was related to anastomosis stenosis in one and obstruction at location of the venous cross clamp canula in three patients. Additionally, complete occlusion of the innominate vein necessitating recanalization in two patients was managed with telescope technique followed by a BeGraft peripheral covered stent (Bentley).

External compression by the Norwood or DKS was the most common cause of LPA stenosis. This is best explained by the small retro-aortic space after surgery. The distance between the pulsatile neo- and descending aorta in the region containing the LPA and left bronchus is small, resulting in anterior-posterior or cranial external compression of the PA branch [4,6,7]. Stenting seems the best option to treat this kind of stenosis, as the stenosis is inaccessible or unsuitable for surgery and balloon dilation is not effective. Though, the forces of a stent on surrounding structures, in particular the airway, in this close interacting trias should be taken into consideration as compression can occur [8,16]. Other causes of PA stenosis were surgical clips, hypoplasia, anastomosis stenosis, post-surgical stenosis and position of the initial PA bifurcation. 3DRA is beneficial to not only visualize the cause of stenosis, but also to visualize the interactions between PA and bronchus or coronary arteries. Furthermore 3DRA provides optimal angulation for intervention as well as 3D roadmapping. In this series, an extra 3DRA run (with balloon interrogation) was used to assess procedural safety in relation to coronary arteries or bronchus in 19 patients (40%). Such an evaluation cannot be performed prior to catheterization and has to be performed during the procedure.

Most stenting procedures were performed for pressure gradients with aberrant vessel morphology. Though, in ten patients no pressure gradient was found and stenting was performed for either aberrant vessel morphology or on clinical indication. Three of these patients were palliated with a bilateral cavopulmonary connection and three other had multiple large collaterals. In those situations the mean cavopulmonary pressure is lower and gradients are often absent, making these gradients unreliable.

Different types of stents are available for the treatment of vessel stenosis in congenital heart disease [3]. For TCPC stenosis Palmaz Genesis stents [5,6,10,14,17,18] and CP stents [10,19] were often used, though we mostly use EV3 and Cook stents that are less applied by others [5,6,17,18]. Our preference for these two types of stents comes from the possibility to insert those stents in small patients (5 kg) at a young age (3 months) with small sheaths (5-6Fr), while maintaining the opportunity to redilate these stents to an adult size of 16 mm for pulmonary arteries [20]. By performing stent interventions this early in life, the problem of vessel stenosis is tackled in a crucial phase of vessel development promoting growth [4]. Besides, these stents give little intima proliferation and thus less restenosis. This approach proved to be successful and safe relieving vessel or conduit stenosis in all patients with an adverse event rate of 10.2%, lower than reported for catheter-based interventions [15,21]. Three patients suffered from thrombus formation as a major adverse event, which were most likely the result of insufficient heparin therapy. Our experience taught us that in particular small patients, patients with central lines and patients with chronic heparin therapy are prone to thrombus formation, despite the per protocol use of heparin 100IE/kg. In two cases an ACT test was performed and was $<$200 s. Because of these results we introduced standard ACT testing during catheterization. An ACT $>$200 s is defined as sufficient and the ACT is performed every 30 min during the procedure. No thrombotic complications were seen after introduction of ACT testing.

In a few patients covered stents (CP, Bentley or ADVANTA) were used. Reasons were reduction of the risk of friction or erosion by an adjacent stent and elective in cases of tunnel stenosis, a fresh thrombus or complete vessel occlusion necessitating recanalization.

Stent fractures were not observed. This is best explained by fact that stents are placed in low pressure areas where there is no pulsatile flow or friction, which lowers the risk of fracturing. Furthermore the EV3 and Cook Formula have low fracture rates.

4.2. Reinterventions

Recatheterizations were performed in more than half of the patients (27/48). In total 21 patients (43.8%) were free of reintervention of their placed stents during the study period. Of the patients that underwent recatheterization four patients had no intervention and two had an intervention on another location. In the other 21 patients 27 reinterventions were performed, including two PA patch plasties. All reinterventions, with the exception of two (7.4%), were planned: serial stent dilation for somatic growth (63.0%) and stent elongation (22.2%). Median time to reintervention was 1.40 years (0.14–6.10) for serial balloon dilation and 1.13 years (0.94–6.01) for extra stent placement. These results are slightly higher than reported by Noonan et al. but our population was stented at a younger age and earlier stage. In their study of LPA stenosis in single ventricle, patients were stented prior or after TCPC at a median age of 4.7 years (1.3–15.2) and had a median follow-up of 4.2 years (0.5–13.4) versus 3.6 years (0.0–17.2) 4.7 years (0.7–9.5) in our study, respectively [6]. In addition, the recatheterization rate in Noonan

et al. was 74.7%, serial redilation rate 42.5% and stent elongation rate 6.4% after a median period of 2.6 years (0.6–7.5) [6]. The one patient with unplanned reinterventions in our cohort, palliated with a bilateral cavopulmonary connection, suffered from intima proliferation in one of the stents and vessel stenosis between two stents 1 year after stent placement. Balloon dilation for in-stent intima proliferation and extra stent placement to bridge the unstented stenotic vessel was performed, emphasizing the necessity of connecting stents. Overall, in-stent stenosis by intima proliferation was rare (1/48 patients), but serial redilations to match stent diameter with patient growth to an adult size were necessary in 30% of the patients.

4.3. Clinical outcome

During follow-up four patients died (8.3%) after a complicated post-operative course not directly related to their first 3DRA stenting procedure. Two patients with pre-existing PLE still have symptoms and are on medication. In the other patients two developed or relapsed from PLE of which one is currently free of symptoms. The other patients were doing well, with remission of PLE and plastic bronchitis in three.

4.4. Study limitations

This study is limited by its retrospective, single center and non-randomized design. Furthermore, the studied population has a small sample size and a median follow-up of 4.7 years (0.7–9.5).

5. Conclusions

TCPC stenoses, especially of the LPA, are common in children with single ventricle physiology. 3DRA visualizes external compression as one of the main causes of stenosis and is used to safely guide stent interventions. The intraprocedural knowledge of interaction with other intrathoracic structures, e.g. aorta, coronary arteries and airway, is taken into account and enhances procedural safety. Stents can be placed in small children, tackling the problem of vessel stenosis in an early and crucial phase of vessel development with good results in both short and long term. Restenosis by intima proliferation is rare, but serial redilations are necessary to match stent diameter with patient size to an adult size.

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Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: Dr. Krings is a member of the Siemens Advisory Board and a consultant for Edwards LifeSciences. The other authors report no relationships that could be construed as a conflict of interest.

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