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Transcatheter left atrial decompression in patients with dilated cardiomyopathy: bridging to cardiac transplantation or recovery

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Abstract

Background: Left atrial congestion results from backward failure in dilated cardiomyopathy. We aimed to evaluate feasibility and efficacy of percutaneous atrioseptostomy to create a restrictive atrial septum defect in management of dilated cardiomyopathy. *Methods and results:* From June 2009 to December 2016, 27 interventions comprised left atria decompressions in 22 dilated cardiomyopathy patients; 9 females; age: 24 days to 36.9 years; weight: 3–50 kg; NYHA-/Ross class IV (n = 16). Mean left ventricular ejection fraction was 21.5 ± 9.7% and brain natriuretic peptide was 2291 ± 1992 pg/ml. Dilated cardiomyopathy was classified as chronic (n = 9); acute (n = 1) myocarditis; idiopathic (n = 5); left ventricular non-compaction (n = 4); mitochondriopathy, pacemaker induced, and arrhythmogenic (n = 3). Atrioseptostomy was concomitantly performed with myocardial biopsies 6.5 days (±11.7) after admission (n = 11). Trans-septal puncture was used in 18 patients; foramen ovale dilatation was done in four patients. Mean balloon size was 11 mm (range 7–14 mm); total procedure time was 133 ± 38 minutes. No procedural complications were observed. Mean left atrial pressure decreased from 15.8 ± 6.8 to 12.2 ± 4.8 mmHg (p = 0.005), left/right atrial pressure gradient from 9.6 ± 5.6 to 5 ± 3.5 mmHg; brain natriuretic peptide (n = 18) decreased from 1968 ± 1606 to 830 ± 1083 pg/ml (p = 0.01). One patient unsuitable for heart transplantation died at home despite additionally performed pulmonary artery banding and three further left atrial decompressions; five patients were bridged to transplantation, two died afterwards. Functional recovery occurred in the remaining 14 patients and in six after additional pulmonary artery banding. No patient required assist device. *Conclusions:* Percutaneous left atrial decompression is an age-independent, effective palliation treating patients with dilated cardiomyopathy.

Dilated cardiomyopathy is the most common cardiomyopathy and remains the prevailing cause for cardiac transplantation in children and adults.[1,2] The phenotype of dilated cardiomyopathy in children is more heterogeneous than in adults. However, in previous Pediatric Cardiomyopathy Registries, aetiologies could not be determined in over half of cases.[2] Morbidity and, in particular, mortality remain unacceptably high; 5-year rate of death or paediatric cardiac transplantation is reported to be 40–60%.[2,3] Dilated cardiomyopathy is characterised by left ventricular dilation, systolic dysfunction, and secondary diastolic interference.[3,4] Left atrial congestion and hypertension occurs as a result of left ventricular backward failure and mainly related to diastolic dysfunction.[5] Symptoms of tachypnoea and dyspnoea correlate with pulmonary congestion and oedema, negative atrial-ventricular coupling, and atrial tachyarrhythmia.[2,3,5] Diuretic use is largely aimed at reducing ventricular pre-load and treating pulmonary interstitial oedema; it is widely subscribed to as a first line management option, neglecting the side effects.[6,7] If pharmacological treatment fails, the patient's condition is classified as "end-stage".[8] For such patients in affluent countries, referral for transplant may be made, with some requiring the bridge of mechanical support.[6,9] However, the mortality of patients awaiting cardiac transplantation is inversely age-related.[10] This inverse relationship also holds true for the chance of ventricular regeneration and recovery.[11,12] We hypothesise in the setting of left atrial and consecutive pulmonary congestion requiring high-dose or long-term diuretic treatment that atrial decompression should be considered, especially before extracorporeal life support becomes necessary.[7] Our aim was to evaluate the feasibility and efficacy of percutaneous atrioseptostomy for creating a restrictive atrial septal defect, thus

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Table 1. Characteristics, presentation at admission, and outcome of patients diagnosed with several causes of dilated cardiomyopathy.

Pt.	DCM-diagnosis	Gender f/m	Age (days)	Weight (kg)	Height (cm)	F.–Class (I–IV) NYHA Ross	BNP (pg/ml)	LV-EF pre (%)	LVEDD pre (mm)	LVEDD pre z-score	Outcome
1	Chronic myocarditis	m	5626	41.8	158	IV	2298	23	73	5.2	HTX/A
2	Acute myocarditis	m	795	14.0	92	IV	4349	17	45	3.2	A
3	LV-NC	m	847	8.5	82	IV	756	20	48	5.8	A/PAB +
4	Chronic myocarditis	f	1322	10.8	91	IV	1149	21	50	5.1	A/PAB +
5	Idiopathic	f	125	6.4	63	IV	532	12	49	5.7	A
6	LV-NC	m	355	7.2	70	III	387	18	38	4.5	A/HTX
7*	Chronic myocarditis	f	13474	50.4	165	IV	269	40	62	4.4	A
8	LV-NC	m	261	7.3	71	IV	257	29	40	4.9	A/PAB
9	Chronic myocarditis	f	708	12.0	94	IV	4928	27	44	3.5	A*
10	Idiopathic	f	1412	13.9	101	IV	1325	35	56	5.3	HTX/D
11	Idiopathic	m	1339	11.9	90	III	3879	43	40	2.7	A
12	PM-CM	f	2458	19.5	119	II	1138	31	60	4.7	A
13*	LV-NC	f	1207	12.9	98	III	188	20	66	7.2	A/PAB +
14***	Idiopathic	m	276	7.0	71	IV	3270	17	44	5.9	PAB + /D
15	Arrhythmogenic	m	42	5.0	58	III	2937	16	42	7.0	A
16	Chronic myocarditis	m	836	15.4	96	IV	1147	19	52	4.5	A/PAB
17	Idiopathic	f	445	9.0	80	IV	5424	11	52	6.5	A/PAB
18	Chronic Myocarditis	m	205	7.7	76	IV	7991	8	50	6.7	HTX/A
19	Chronic Myocarditis	m	946	11.1	94	III	825	28	42	3.2	A
20	Mitochondriopathy	f	187	6.0	69	IV	3263	4	50	7.7	A/PAB

21	Chronic Myocarditis	m	24	3.1	52	IV	1826	20	26	3.5	A/Pott + bPAB/HTX
22	Chronic Myocarditis	m	82	3.1	50	IV	2261	13	45	9.1	HTX/D
	Mean		1302.2	12.9	88.2	3.73	2290.9	21.5	48.8	5.3	
	Median		708.0	9.9	86.0	4.00	1575.5	20.0	48.5	5.2	
	SD (+/–)		2785.9	11.3	28.6	0.55	1992.1	9.7	10.0	1.6	
	Min.		24	3.1	50	2	188	4	26	2.7	
	Max.		13474	50.4	165	4	7991	43	73	9.1	
	Quartile 25		196				700				
	50		708				1576				
	75		1265				3422				

A = alive; A* = lost to follow-up; BNP = brain natriuretic peptide; bPAB = bilateral pulmonary banding; D = dead; DCM = dilated cardiomyopathy; EF = ejection fraction; HTX = heart transplantation; LVEDD = left ventricular end-diastolic dimension; LV = left ventricular; LV-NC = left ventricular non-compaction; PAB = pulmonary artery banding; PAB+ = pulmonary artery banding + open heart surgery; Potts+ = reverse Potts-Shunt

*One

**Two

***Three additional interventions

unloading the left atrium and ventricle in dilated cardiomyopathy patients with reduced ejection fraction.

Methods

This retrospective study encompasses 27 left atrial decompressions in 22 patients with dilated cardiomyopathy. Patient's median age was 2 years (24 days to 36.9 years), median body weight 9.9 kg (3–50 kg); nine patients were female. The demographic data are summarised in Table 1. According to the Pediatric Cardiomyopathy Registry,[4] dilated cardiomyopathy was defined by depressed left ventricular function with ejection fraction <45% and left ventricular dilatation – left ventricular end-diastolic dimension, z-score >+2. According to the results of histopathological/immunohistological analysis of the obtained endomyocardial biopsies and preferential MRI, 10 patients (45%) revealed dilated cardiomyopathy with inflammation and classified as chronic (n = 9) and acute (n = 1) myocarditis. Chronic myocarditis is histologically defined as co-existence of already degenerated muscle tissue and fibrosis together with still persistence of infiltration of interstitial tissues. The others were classified as idiopathic (n = 5); left ventricular noncompaction (n = 4); or associated with mitochondriopathy, arrhythmias, and pacemaker (n = 3). NYHA/Ross function was classified as class IV (n = 16), III (n = 5), and II for the patient with pacemaker-induced cardiomyopathy. Mean left ventricular ejection fraction was 21.5 ± 9.7%; the left ventricular end-diastolic dimension was 49 ± 10 (z-score 5.3 ± 1.6); median serum brain natriuretic peptide was 1576 pg/ml in the range 188–7991 pg/ml. Percutaneous trans-septal decompression was performed 8.4 (±11.3) days after admission. In 11 patients, concomitant myocardial biopsies were taken. Institutional Review Board approval was obtained for analysing all retrospective data. Informed written consent was obtained before the procedures from all patients or their parents.

Statistical data were calculated with SPSS (IBM). Descriptive data are presented as mean ± SD or median and range, as appropriate. Significance was calculated with the Student's t-test for paired samples. A p-value ⩽0.05 was considered statistically significant. Continuous variables are presented as mean ± SD or median and range as appropriate. The calculated z-score is given for the age-dependent left ventricular end-diastolic dimension; a z-value of +2.5 was defined as dilated cardiomyopathy. Specific parameters were analysed only if both pre- and post-intervention data were available.

Interventional technique

All procedures were performed under fluoroscopy guidance and only by local anaesthesia or in young children with analgosedation (Fig 1a–d). No patient underwent general anaesthesia; all received local anaesthetic. In stable patients a complete haemodynamic assessment was performed before intervention. In critically unwell patients, the intervention was performed almost in isolation. In n = 18, MRI was performed before or as a hybrid imaging procedure with subsequent heart catheterisation. In younger patients, especially infants, this was carried out under a single episode of sedation; the total procedure time is presented in Table 2. Trans-septal puncture was performed in 18 patients using a Brockenbrough technique (Table 2) with a needle length of 56 or 71 cm (Cook-Medical). In children, the needle was loaded in a 6F, 48 cm-long Check-Flow performer® Introducer (Cook-Medical); in adolescents and adults an 8F Mullins sheath

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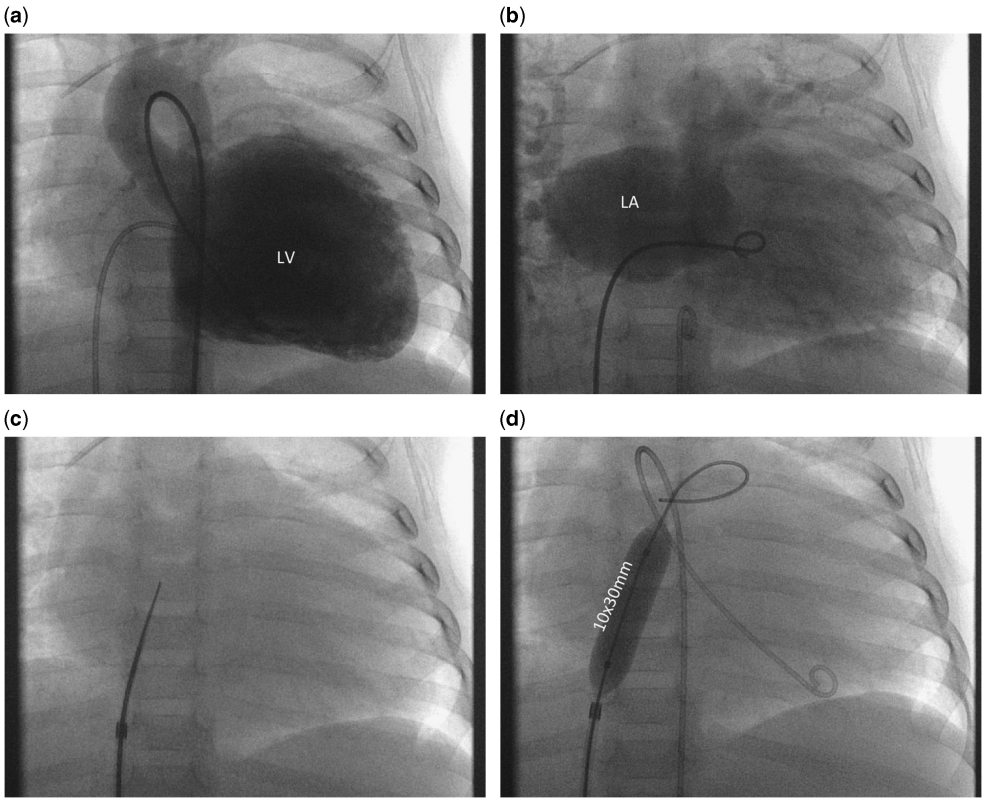

Figure 1. (*a–d*) Left ventricular (LV) dilated cardiomyopathy of patient no. 17 at day 445, when a restrictive atrial communication was created. On (*a*) the left ventricular angiography with an extreme dilated LV is depicted (MRI; z-score of left ventricular end-diastolic dimension was 6.5). (*b*) Angiographically obtained left atrial dimension before the atrial septum puncture. (*c*) Trans-septal needle advanced through a 6F Cook sheath in anterior–posterior plane; before the atrial septum is punctured the correct posterior direction to the left atrium has additionally to be demonstrated in the 90° lateral frame (see text); (*d*) Final Powerflex® balloon fully inflated to a diameter of 10 mm within the atrial septum. A diagnostic Pigtail catheter is still re-advanced in the LV cavity.

(Cook-Medical) was used. To ensure safety during manipulation the long sheath was advanced through the already placed 2F larger short sheath. The needle-sheath ensemble was pulled back from the upper caval vein in the right atrium during continuous application of contrast medium through the needle. For interatrial septum puncture, the tip of the puncture ensemble was directed posteriorly on the 90° lateral view and between a 3 and 4 o'clock positions on anterior–posterior view. With contact of the fossa ovalis the needle was advanced to inject contrast medium into the atrial septum, then advanced to the left atrium by further continuous injection of contrast medium. After entering the left atrium, the Cook sheath was advanced across the septum and the needle replaced with a 0.035 in. guidewire. Balloon dilatation of the atrial septum was then performed with high-pressure balloons; in most, depending on the patient's size, Powerflex® (Cordis, Switzerland) or Atlas® (Bard, United States of America) balloons with a length of 20–40 mm were used. The maximal balloon diameter was chosen depending on the desired size of the atrial communication. In all patients, the goal was to achieve a restrictive atrial communication; therefore, a somatic size-dependent communication between almost 4 and 8 mm was created. Based on our long-term experience,[8] static ballooning of the intact atrial septum was in part performed by sequential dilatation using balloon diameters up to a maximum of 14 mm resulting in an arterial communication of up to 8 mm. Powerflex® 12 × 30 mm balloons or, in adolescents and adult patients, Atlas® balloons (14 × 20 mm) were predominantly used as the final balloon diameter with an average balloon diameter of 11 mm (min 6 mm, max 14 mm) (Table 2). Indentation by the atrial septum on the waist of the inflated balloon, followed by the disappearance of the waist after full inflation was identified in all procedures (Fig 1a–d). If recoil was observed during deflation, repetition of inflation up to the rated burst pressures (8–18 atm.) was performed. At the end of the procedure, full or only partial haemodynamic data were obtained and the created atrial communication was evaluated by transthoracic echocardiography. Persistent foramen ovale dilatation without trans-septal needle puncture was performed in four patients. The same balloon material and technique was used as described. In one patient, an Andra-stent XL was placed into the intact atrial septum, mounted on a 12 × 30 mm balloon and inflated to a residual diabolo shape, with a waist diameter of 8 mm. Routine stenting of the atrial septum was avoided, to reduce additional procedural risks and to prevent negative aspects of long-term stent placement considering also the chance for recovering. Expecting clinical and haemodynamic improvement after creation of an atrial defect, intended right ventricular myocardial biopsies were always performed after generation of the restrictive septum defect. Procedural success was defined as a successful creation of a patent, but restrictive atrial communication. Restriction was defined by echocardiography diameters between 4 and 10 mm and/or residual left/right atrial pressure gradient of >3–5 mmHg. Gradual balloon dilatation of the intact atrial septum was favoured in some patients, to take care and attention not to create an unrestrictive atrial communication. The decision for re-dilatation of the atrial septum during the later follow-up was made depending on clinical

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Table 2. Procedure related data including the left atrial pressure (LAP) before and after balloon dilatation of the atrial septum, in 18 patients after trans-septal puncture by Brockenbrough technique and 11 additional myocardial biopsies during the same heart catheterisation (y+, cath.).

Patient	LAP-pre (mmHg)	Brockenbrough (yes/no)	Procedure time (min)	Fluoroscopy time (min)	Balloon size max. (mm)	LAP-post (mmHg)	Myocardial biopsy y+ = same Cath.
1	26	No (ballooning)	143	23.1	12	17	y
2	23	Yes	148	21.2	14	18	y+
3	17	Yes	193	21.1	12	15	y
4	22	Yes	143	12.2	12	19	no
5	9	Yes	108	14.3	10		y
6	17	Yes	106	12.0	10	9	no
7*	9	Yes + stent	82	7.4	14	7	no
8	15	Yes	142	18.0	10	10	no
9	33	no (ballooning)	122	19.4	12	20	y+
10	23	Yes	102	15.1	10	19	no
11	13	Yes	210	34.0	10	12	y+
12	9	Yes	185	25.3	12	9	y+
13*	14	Yes	159	19.3	10	6	no
14***	12	No (ballooning)	90	13.2	7	10	no
15	7	No (ballooning)	127	9.3	7	7	y+
16	11	Yes	165	26.0	10	8	y+
17	9	Yes	112	15.4	10	10	y+
18	24	Yes	92	17.5	10	21	y+
19	17	Yes	173	14.1	10	10	y+
20	11	Yes	57	8.2	8	9	y+
21		Yes	126	6.1	12	13	y
22	12	Yes	147	17.2	10	8	y+
Mean	16		133	16.8	11	12	
Median	14		135	16.3	10	10	
SD (+/−)	7		38	6.6	2	5	
Min	7		57	6.1	7	6	
Max	33		210	34.0	14	21	

Cath. = catheterisation; LAP = left atrial pressure

symptoms of pulmonary congestion and echocardiographic evidence of recoil of the atrial septum causing an unwanted degree of restriction at the created atrial communication.

Results

Creation of a restrictive atrial communication was successful in all patients. There were no procedural death or complications. Dilated cardiomyopathy remained idiopathic in 23% of the patients. The average mean procedural length, including MRI, haemodynamic assessments, and myocardial biopsies, was 133 ±38 minutes; median fluoroscopy time 16.8 minutes (range 6.1 and 34 minutes) (Table 2). Mean left atrial pressure pre-intervention decreased from 16 ± 7 before atrioseptostomy to 12 ± 5 mmHg post-intervention ($p = 0.005$). The left/right atrial pressure gradient decreased from 9.6 ± 5.6 to 5 ± 3.5 mmHg ($p = 0.005$). Left atrial and left ventricular end-diastolic pressure was recorded in seven patients; in this group, both pressures decreased by a mean of 2 ± 1 mmHg. The left ventricular ejection fraction at discharge ($n = 18$) increased significantly from 21.5 ± 9.7 to 29.2 ± 8% ($p = 0.01$). This correlated with immediate improvement of NYHA/Ross functional class from a mean of 3.73 (0.55 ±) to 2.95 (±0.84). Serum brain natriuretic peptide ($n = 18$) decreased from median 1576 pg/ml (range: 188–7991 pg/ml) to

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418 pg/ml (range: 89–4349 pg/ml) ($p = 0.05$) between admission and discharge.

Follow-up

The median in-hospital followed-up time of all treated patients was 42 (range 1–253) days; outcome ($n = 22$) was followed over a median time of 528 (2–2262) days after the procedure. Following the primary intervention, three patients received a second intervention 27, 337, and 645 days later. In patient number 14, the atrial septum was manipulated four times within 71 days. Re-interventions were clinically and echocardiographically indicated for re-dilatation of patient, but recoiling and increasingly restrictive atrial communication. After creation of a restrictive atrial communication all patients improved; three patients with the need for re-dilation only transient. Patient 14 requiring four atrial manipulations and additional pulmonary artery banding died finally 225 days after the first intervention; the family had declined cardiac transplantation. Six patients, who remained listed for cardiac transplantation, were successfully transplanted; two of them died 7 and 13 months after transplantation because of acute graft failure, one associated with a relapse of a fulminant cytomegalovirus and liver failure; the other patient related to probable parental compliance problems.

One patient survived pulmonary artery banding procedure, but without functional cardiac improvement until the end of the observation period; all others showed functional recovery and regeneration with normalisation of left ventricular end-diastolic dimension, six patients, though one recovered after additional pulmonary artery banding placement. Three patients received pulmonary artery banding after open-heart surgery before weaning from cardiopulmonary bypass. Re-shifting of the interventricular septum to the left was related to a consecutive change of the left ventricular cavum from a globated to an ellipsoid form. The decision for pulmonary artery banding was made according to our previously published inclusion criteria,[13] which provides a normal right heart function with still normal right atrial and end-diastolic pressures and a pulmonary artery pressure of less than half of the systemic blood pressure level. The final functional class of all, but one survivor was class I in $n = 10$ patients, II ($n = 7$), and III ($n = 1$), respectively.

Discussion

This retrospective study reports the impact of transcatheter creation of a restrictive atrial communication in heterogeneous causes of patients with dilated cardiomyopathy. Preliminary human studies have proposed left atrial dysfunction in heart failure with preserved ejection fraction; creation of an interatrial communication was described as a novel strategy.[14,15] The majority of reports are focussed on left atrial decompression during "extra-corporal life support".[16–18] Creation of an interatrial communication in patients with reduced left ventricular ejection fraction is currently not reported. However, the here-described trans-septal needle technique followed by static balloon dilation of the atrial septum is safe and effective regardless of patient's age, clinical condition, or cause of cardiomyopathy. At our institution, atrioseptostomy with the described technique is established since newborns with hypoplastic left heart syndrome are treated by "Giessen hybrid" approach.[19,20] Based on our long-term experience of atrioseptostomy, the difference of a fully unrestricted atrial communication and the pathophysiological consequences of an overly restrictive atrial septum defect on the haemodynamics and pre-load-dependent performance of the systemic ventricle are previously described.[21] In newborns and infants with borderline left heart structures, the importance of adequate pre-load on left heart growth properties is evident.[21] Considering the pathophysiological features of congestive heart failure, it is natural that restrictive ventricular filling carries prognostic implications in dilated cardiomyopathy patients.[5,22] We hypothesised that atrial and consecutive pulmonary congestion can be effectively influenced if a left ventricular pre-load is optimised to reduce congestive symptoms, but still permits an adequate left ventricular filling. We note that all the reported patients in this study benefitted from creation of a restrictive atrial communication in both echocardiographic and, importantly, clinical findings. However, haemodynamic and clinical improvements did not necessarily correlate with a change of shape and form of the affected systemic ventricle. Therefore, in whom the criteria for pulmonary artery banding, such as age, right ventricular affection, pulmonary arterial hypertension, written parental consent, with or without an additional open-heart surgery were not fulfilled or spontaneous improvement could not be observed, the decision to remain the patients listed for transplant was based on missing alternatives forcing cardiac regeneration or because of progressive heart failure despite generated restrictive atrial septum defect. Naturally, the most severely impaired patients with acute or chronic left atrial hypertension and consecutive pulmonary congestion showed the most beneficial effect. Patient 14 can exemplarily best demonstrate this; following the declination of cardiac transplantation by the family, three repeat atrial septum interventions were performed during the follow-up, each time the infant's clinical condition improved and discharge from hospital became possible. Contemporaneously, this case also shows the weakness of exclusive static balloon dilatation for creating an atrial communication. Considering the variable anatomical structure of the atrial septum, the long-term efficacy of the created communication solely performed by balloon dilatation is unpredictable. However, stent placement was avoided as a primary approach as mentioned earlier; flow reduced devices might have a further option, if they become available. In addition, it should be remarked that carefully performed gradual atrial septum balloon dilatation, using sequentially increased balloon diameters, avoids creation of unrestricted atrial septum defects. The function of a restrictive "pop-off valve" does not lead to right ventricular volume overload, but adequate unloading for the failing left ventricle, which could be demonstrated by the patient's clinical functional class at rest and exercise. Empirically and according to our previous institutional experience[8,21] as well as based on computer modelling,[23] an atrial communication of 4–10 mm remains – depending on the patient's somatic-size – restrictive and serves as an acute or chronic "pop-off valve" reducing cardiac congestive symptoms. Left atrial decompression with a physiologic pressure gradient between the right and a left atrium of about 3–5 mmHg provides adequate left atrial decompression such that an immediate anti-congestive effect was observed in this cohort of patients with reduced left ventricular ejection fraction and increased left ventricular end-diastolic dimension. Concomitant with a significant decrease of the left atrial pressure, the left atrioventricular coupling improved, which could be demonstrated in those seven patients in whom complete pre- and post-intervention haemodynamic data were obtained. Left ventricular end-diastolic pressure decreased in line with reduction of left atrial pressure.

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Theoretically, decreased left atrial pre-load with subsequent decrease in left ventricular filling pressure causes a shift within the Frank-Starling curve, which might support favourable remodelling processes, as seen in some of our especially young dilated cardiomyopathy patients. Further prospective studies must clarify whether creation of a restrictive atrial communication contributes really to left ventricular functional recovery though the theoretical advantages are reduced left ventricular wall stress with secondary improvement in sub-endocardial perfusion and reduced myocardial oxygen consumption. Considering our heterogeneous causes of patients with dilated cardiomyopathy, including a high variability of age, it seems that not the disease itself but the consecutive clinical condition and pathophysiology should determine the indication for generation of a restrictive atrial communication. Pathological haemodynamics, including left atrial size and function with the need of diuretic treatment, are the main consideration of establishing a restrictive atrial communication. Based on our further interventional and surgical experience dealing with all faceted congenital and acquired heart failure, we propose that the presence of a restrictive atrial communication is almost always of haemodynamic and clinical advantage even in patients with reduced ejection fraction. Therefore, maintaining or creating of a de novo restrictive atrial communication belongs to our holistic strategy in the management of patients with atrial and pulmonary congestion based also on a reduced left ventricular ejection fraction with increased end-diastolic dimension.[8] Despite the limitations of this retrospective analysis, the reported results are promising. Bridging to recovery or transplantation with a less incidence for need of mechanical assist devices are realistic scenarios as well as improved quality of life corresponding with improved haemodynamic parameters and biomarkers. However, the results need to be confirmed further by prospective multi-centre studies. But ascertainable is that infants and children have the best potential for recovery and remodelling, if novel interventional and surgical procedures are combined with adequate age-dependent medical heart failure therapy.[23,24] The current report demonstrates a sustained and relevant clinical benefit of a restrictive atrial septum defect as a part of a holistic therapeutic approach. In addition to the efficacy, there was no need for closure of the created atrial defects during the further follow-up.

Study limitations

The study is limited by its retrospective design and partially incomplete data, in particular considering concomitant diastolic dysfunctional data. Furthermore, follow-up cardiac MRI data was not available to better describe the impact of intervention. We were unable to present all detailed haemodynamic measurements and close follow-up. The described cohort is small and heterogeneous in respect to age, body size, and underlying causes of dilated cardiomyopathy. Moreover, there were several different surgical interventions to several patients during the follow-up period. Furthermore, we cannot offer evidence of all underlying pathophysiological or molecular mechanisms that may underpin the clinical findings. Missing data may affect bias and thus, affect the validity of the statistical analysis.

Conclusion

Our experience in creation of a restrictive atrial communication in a small cohort of children and young adults with symptomatic dilated cardiomyopathy is encouraging and might be the basis for prospective multi-centre studies. In patients with reduced left ventricular ejection fraction, decompression of the left atrium by creation of a restrictive atrial communication should be considered as a safe, cost-effective palliation, when symptoms of pulmonary congestion or intrinsic left atrial hypertension become obvious. We are convinced that unloading of the left atrium should be considered, when high dose or long-term diuretic treatment with negative anti-remodelling effects becomes necessary.[25,26] In our experience thus far, creation of a restrictive atrial septum defect in patients with left ventricular ejection fraction with increased end-diastolic dimension is both, safe and effective. Indications for the procedure must not be limited to clinicians with access to inter-atrial flow regulator devices;[27] rather, this important intervention must be made available to all patients who may benefit.

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Conflicts of interest. None of the authors have anything to disclosure.

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