Growth of hypoplastic mitral valves in hypoplastic left heart complex and similar constellations after anatomical left superior vena cava correction

Robert A. Cesnjevar, Frank Harig, Moritz Dietz, Muhannad Alkassar, Wolfgang Waellisch, André Rueffer, Sven Dittrich and Ariawan Purbojo

Department of Pediatric Cardiac Surgery, University Hospital Erlangen, Erlangen, Germany
Department of Pediatric Cardiology, University Hospital Erlangen, Erlangen, Germany

* Corresponding author. Kinderherzchirurgische Abteilung, Universitätsklinik Erlangen, Loschgestrasse 15, 91054 Erlangen, Germany. Tel: +49-9131-8534010; fax: +49-9131-8534011; e-mail: robert.cesnjevar@uk-erlangen.de (R.A. Cesnjevar).

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Abstract

OBJECTIVES: Left superior vena cava (LSVC)-related obstruction of mitral inflow is a rare finding in patients with complex cardiac anomalies like hypoplastic left heart complex. We report our experience by establishing a left superior to right superior caval vein continuity (innominate vein creation by direct LSVC–right superior vena cava end-to-side-anastomosis), and coronary sinus unroofing if indicated for LSVC-related mitral inflow obstruction.
METHODS: Nineteen patients (median age: 1.0 ± 0.3 years; range: 7 days–4.8 years) underwent anatomical correction of LSVC without the use of foreign material in conjunction with repair or palliation of congenital anomalies in a single centre between April 2015 and November 2019. Indications for the procedure were LSVC-related obstruction of left ventricular inflow due to a dilated coronary sinus. Additional procedures included mitral (n = 7) or atroioventricular (n = 3) valve surgery, right ventricular to pulmonary artery conduit (n = 3), first stage palliation (n = 3) or biventricular repair (n = 5) of hypoplastic left heart complex. Three patients needed secondary mitral valve replacement (n = 3).

RESULTS: All LSVC or coronary sinus-related obstructions were effectively relieved. No patient died early, 2 patients died later after the procedure. One patient needed stenting of the superior vena cava below the unobstructed cephalad vein anastomosis at the former right superior vena cava-cannulation-site. Follow-up was complete and demonstrated an 89.5% survival after 2.5 ± 0.4 years. Innominate vein patency was 100% documented by echocardiography (n = 19), cardiac catheterization (n = 6) or both. Mean mitral valve z-scores before the operation were -1.7 ± 0.2 (range -3.8 to 0.3) and increased to 0.7 ± 0.2 (range -0.7 to 1.9) after LSVC repair.

CONCLUSIONS: Anatomical correction by surgical creation of an innominate vein is an effective method to relieve LSVC-related obstructions and promotes mitral valvar growth. Mitral ring sizes were at least normalized after surgery at the time of discharge. Further prospective follow-up studies to evaluate the growth potential of left-sided heart structures by reporting cardiac z-scores are needed to evaluate the true impact of coronary sinus unroofing.

Keywords: Left superior vena cava • Hypoplastic left heart complex • Mitral valve growth • Mitral valve z-score • Congenital heart disease

ABBREVIATIONS

| Abbreviation | Description                      |
|--------------|----------------------------------|
| HLHC         | Hypoplastic left heart complex    |
| LSVC         | Left superior vena cava          |
| PLSVC        | Persistent left superior vena cava|
| RSVC         | Right superior vena cava         |
| VSD          | Ventricular septal defect        |

INTRODUCTION

A persistent left superior vena cava (PLSVC) is an embryological venous remnant of a cardinal vein occurring in 0.3% of the healthy population and in 1.3–11% of patients with cardiac anomalies [1–4]. The presence of a left superior vena cava (LSVC) during surgery is sometimes annoying for cardiopulmonary bypass because achieving adequate venous drainage may be more difficult. But in general, it is supposed to be a rather irrelevant anatomical feature with absolutely no pathophysiological importance. However, some authors have emphasized that PLSVC may be of more concern as it is often present in patients with left-sided obstructive lesions creating left ventricular inflow obstruction to some extent [4–6].

The normal LSVC collects the left jugular, subclavian and hemiazygos veins before it enters the left atrium close to the left atrial appendage. The normal course as a ‘closed venous tube’ is through the left atrium in front of the posterior mitral valve annulus, before it drains into the right atrium at the level of the coronary sinus [4]. There have been several anecdotal reports on surgical procedures on how to address especially large-sized LSVCs as they may have some impact on mitral valve inflow. A large venous ‘hump’ in front of the mitral annulus has been reported to cause relevant supravalvar mitral stenosis with congestion [1, 5–14]. The anatomical proximity to the mitral annulus makes such a pathophysiological impact plausible. Cochrane et al. reduced the intra-atrial LSVC size with a running suture, Kreutzer et al. ligated the LSVC and connected it to the other side with a Gore-Tex® graft and some authors like Vargas et al. advocated an end-to-end-anastomosis with the right atrial appendage similar to the Warden procedure. However, to date, only some authors have challenged a true anatomical repair by surgically creating an innominate vein with a direct end-to-end-anastomosis of the mobilized LSVC to the right superior vena cava (RSVC), which has become our surgical approach for this condition since 2015 [15, 16].

Reviewing our own experience and the literature brought us to the idea that there might be a more relevant impact of LSVCs for patients with hypoplastic left heart complex (HLHC) or borderline-sized left ventricles. Its permanent, often subclinical mitral inflow obstruction may have a constant negative impact on mitral valve growth [4–6, 10]. Kadletz et al. published a case report from Toronto with a borderline-sized left ventricle and mitral pathology in the presence of a huge LSVC in conjunction with a missing right-sided vena cava. The authors stated in their case report that the presence of a large LSVC is probably responsible for the development of later left ventricular hypoplasia. However, the very special and unique pathology in this patient was the additional absence of an RSVC as well, which may have aggravated the situation.

The hypothesis that a large left-sided vena cava influences LV development by limiting mitral inflow was supported by our first corrected index patient with mitral hypoplasia, a large PLSVC, smallish LV, a mid-muscular ventricular septal defect (VSD), hypoplastic aortic anulus, aortic arch and coarctation, where we repaired the arch obstruction together with pulmonary artery banding and concomitant LSVC repair. The left ventricle and mitral annulus grew in size and the patient underwent successful VSD-closure and debanding as a secondary repair 9 months after the initial operation. The positive initial experience convinced us to investigate the influence of our anatomical surgical LSVC repair systematically with special interest on LV development and mitral valve growth.

PATIENTS AND METHODS

Clinical setup

Nineteen patients entered systematic follow-up after admission for treatment in our centre since 2015. Surgery was indicated by an institutional heart team of congenital cardiac surgeons and
paediatric cardiologists during a preoperative joint cardiac con-
ference (Cardio-board). Before patients were scheduled for sur-
gery, their clinical data and all acquired preoperative diagnostic
images were systematically reviewed. After discharge, patients
were seen by outpatient cardiologists from our institutional net-
work (Netzwerk AHF Nordbayern) and retransferred to our
centre when special imaging (computed tomography/magnetic
resonance imaging) or a diagnostic cardiac catheter became ne-
cessary. The study was approved by our local ethics commit-
tee; data were collected retrospectively from the patients’ surgical
and cardiological hospital records. The retrospective data collect-
ion used electronic imaging software which included measure-
ment tools (syngo®Dynamics-workplace, Siemens-Healthcare®,
Version July 2018) for retrospective imaging reviews. Measurements were performed by 2 experienced paediatric car-
diologists using standardized planes.

Patients’ diagnoses and characteristics are presented in
Table 1.

Surgical procedure

The LSVC was completely mobilized from the left subclavian vein
down to the left atrial appendage in the absence of a bridging
(innominate) vein. Small side branches and the vena hemiazygos
were ligated or clipped. SVC, LSVC and IVC were cannulated after
standard arterial cannulation on the fully heparinized patient.
The LSVC was detached from its atrial connection. The meta-
atrial LSVC part in front of the mitral valve orifice was completely
reseated during cardiopлегic arrest. The mitral valve with its sub-
valvar apparatus was inspected and sized. The mitral valve was
repaired or replaced if necessary. The LSVC was end-to-side
anastomosed to the R SVC with a 7 × 0 running polypropylene
suture (Prolene®, Ethicon) at the end of the procedure during
rewarming of the patient on bypass with the beating heart. The
anastomosis was interrupted on 2 opposite ends to avoid a pos-
sible purse-string effect (see Fig.1). The anastomotic function was
checked by central venous pressure-measurements, near infrared
spectroscopy-monitoring and sequential echocardiographies.
Some patients underwent angiocardiography postoperatively for
suspicion of stenosis (n = 1) or routine postoperative assessments
before the next surgical steps were undertaken (n = 5).

Postoperative anticoagulation

In general, the primary disease triggered postoperative anticoa-
gulation for all patients. The venous anastomosis attached native
tissue to native tissue, so we have not seen any specific indication
for postoperative systemic long-term-anticoagulation. Patients
were routinely heparinized postoperatively with an intravenous
rate of 10 IU/kg/h for neonates and patients below 12 months.
The remaining older patients received 5 IU/kg/h heparin intra-
venously. All patients receiving any kind of mitral valves were
fully heparinized (partial thromboplastin time 50–60 seconds)
postoperatively and switched to Coumadin® afterwards, which
was the case for 1 patient after SVC-stenting as well.

Echocardiography

The patient’s mitral valve size was measured preoperatively by 2-
dimensional-echocardiography (GE Vivid E95 and Vivid 7, GE
Healthcare; Philips Affinity 50G, Royal Philips Healthcare com-
pany) in 2 standard planes (four-chamber and long-axis view)
with the left-sided-AV-valve open in diastole and was re-
evaluated sequentially on regular postoperative visits. Changes
were expressed using z-scores to express mitral valve hypoplasia,
mitrval valve growth and development. Each patient served as his
own control.

Follow-up

Follow-up after surgery was 2.5 ± 0.4 years and complete (100%).
Patients were included in a regular institutional follow-up, as
most of the patients underwent staged repair (n = 12; 63.2%).
Previously, palliated patients had a complete diagnostic workup
(incl. cardiac cath) before definite repair.

Statistical analysis

Statistics were performed using statistical software (EXCEL,
Microsoft and SPSS for Windows, Microsoft Corporation,
Redmond, WA, USA). The data are expressed as mean with a
standard deviation of the mean values (SEM, standard error of
the mean). Significance tests were carried out using Student’s t-
test or the non-parametric U-test, according to the nature of the
tested variables. A statistically significant difference was suspected
to exist at a probability value below 0.05 (P < 0.05).

RESULTS

There was no early mortality, 2 patients died late during follow-
up. One patient died late after failed primary biventricular repair
at the age of 10 months. Postoperative low cardiac output with
multiorgan failure forced us to modify his initial repair by a previ-
ously published rescue procedure with bilateral banding and im-
plantation of a ‘reverse shunt’ (Pott-shunt 6 mm Gore-Tex®) from
the main pulmonary artery to the descending aorta [17]. The se-
cond patient with heterotaxia and pulmonary atresia died
9 months after biventricular repair in an external paediatric hos-
pital due to a fulminant septic event, possibly related to pulmon-
ary valve endocarditis of his RVPA-conduit. Unfortunately, the
parents refused post-mortem autopsy despite the surprisingly
rapid deterioration of their child’s condition after hospital
admission.

Mitrval valve size increased in some patients intraoperatively
immediately after removal of the intracardiac LSVC tubular
course by several millimetres (mean 4.1 ± 0.5 mm, range 1.0 to
8.6 mm). This fact is difficult to understand and explain, as growth
is practically not possible in such a brief period (see Figs 2 and 3).
Three patients underwent mitral valve replacement in a supra-
nnular (n = 2) or intra-annular position (n = 1) with an oversized
prosthetic valve. Both supra-annular implanted valves were over-
sized mechanical valves for older children. One patient received
a biological xenograft twice to replace his severely dysplastic mi-
tral valve at the first operation, which was later replaced with a
Melody valve due to early degeneration. The surgically achieved
mitral valve orifice was defined as the size of the implanted pros-
thesis for these patients.

Mitrval valve size seemed to increase over time for all observed
patients significantly. Starting from a mean z-score of -1.7 ± 0.2,
## Table 1: Patient diagnosis

| Patient | Sex | Age (months) | Diagnosis | Primary repair | Outcome | Status | Patency | Follow-up (years) | RACHS | Aristotle score |
|---------|-----|--------------|-----------|----------------|---------|--------|---------|------------------|--------|-----------------|
| 1.      | M   | 45           | HLHC, Shone-complex, s.p. repair of aortic arch hypoplasia and coarctation, mitral stenosis | No | Biventricular repair MV replacement | Alive | Yes | 3.8 | 4 | 11 |
| 2.      | M   | 5            | Unbalanced AVSD | Yes | Biventricular repair | Alive | Yes | 4.7 | 3 | 9 |
| 3.      | M   | 11           | VSD, mitral stenosis, supravalvular membrane | Yes | Biventricular repair | Alive | Yes | 4.7 | 3 | 8 |
| 4.      | M   | 7            | AVSD and coarctation, smallish LV | Yes | Biventricular repair | Alive | Yes | 4.2 | 3 | 8 |
| 5.      | M   | 57           | AVSD, smallish LV | No | Biventricular repair, MV replacement | Alive | Yes | 3.9 | 6 | 15 |
| 6.      | F   | 0.2          | HLHC, hypoplastic aortic arch, coarctation, mitral stenosis and VSD | No | Biventricular repair | Alive | Yes | 3.3 | 4 | 11 |
| 7.      | M   | 39           | HLHC, aortic atresia, hypoplastic aortic arch and VSD | No | Biventricular repair (Mustard–Rastelli) | Alive | Yes | 2.3 | 4 | 11 |
| 8.      | F   | 4            | Unbalanced AVSD | Yes | Biventricular repair | Alive, stent in RSVC | Yes | 2.3 | 3 | 9 |
| 9.      | F   | 6            | TGA, VSD, PS, smallish LV | Yes | Biventricular repair | Alive | Yes | 2.3 | 4 | 11 |
| 10.     | M   | 39           | AVSD, common atrium, congenital complete heart block | No | Biventricular repair | Alive | Yes | 2.3 | 3 | 9 |
| 11.     | M   | 0.5          | HLHC, hypoplastic aortic arch, hypoplastic aorta, coarctation, mitral stenosis | Yes | Failed biventricular repair converted to reverse shunt (Pott-shunt) and bilateral PA banding | Died before re-conversion to biventricular repair (pneumonia) | Yes | 0.8 | 6 | 15 |
| 12.     | F   | 2            | HLHC, multiple VSDs, Cor triatriatum, mitral hypoplasia, smallish aorta | Yes | Biventricular repair | Alive | Yes | 2.2 | 3 | 8 |
| 13.     | F   | 4            | HLHC, hypoplastic aortic arch, coarctation, hypoplastic aortic valve, mitral hypoplasia | No | Biventricular repair | Alive | Yes | 1.7 | 6 | 15 |
| 14.     | M   | 13           | Heterotaxia, TAPVD, DORV, PA | No | Biventricular repair (Mustard–Rastelli) | Died 1.5 years after biventricular repair | Yes | 0.6 | 4 | 11 |
| 15.     | M   | 0.1          | HLHC, M. Shone, hypoplastic mitral valve, hypoplastic aortic arch and coarctation | No | Biventricular repair, MV replacement and later rereplacement[AQ] | Alive | Yes | 1.5 | 6 | 15 |
| 16.     | F   | 14           | DORV, hypoplastic mitral valve and hypoplastic LV, supravalvular membrane | No | Still awaiting biventricular repair after LSVC rerouting | Alive | Yes | 0.7 | 3 | 8 |
| 16.     | F   | 14           | DORV, hypoplastic mitral valve and hypoplastic LV, supravalvular membrane | No | Still awaiting biventricular repair after LSVC rerouting | Alive | Yes | 0.7 | 3 | 8 |
| 17.     | M   | 11           | VSD, mitral stenosis, Down syndrome | No | Biventricular repair | Alive | Yes | 0.6 | 3 | 8 |
| 18.     | M   | 7            | HLHC, M. Shone, hypoplastic aortic arch, coarctation, hypoplastic mitral valve, supra-mitral membrane | No | Biventricular repair | Alive | Yes | 0.6 | 3 | 8 |
| 19.     | M   | 11           | HLHC, M. Shone, hypoplastic mitral valve, LVOTO, hypoplastic aortic arch, coarctation, art. lusoria | Yes | Biventricular repair | Alive | Yes | 0.4 | 6 | 15 |

Biventricular repair was either VSD repair and mitral reconstruction (RACHS 3, Aristotle score 8), AVSD repair (RACHS 3, Aristotle score 9), repair of complex transposition (RACHS 4, Aristotle score 11), Ross–Konno Operation (RACHS 4, Aristotle score 11) or biventricular repair of HLHC (RACHS 6, Aristotle score 15) resulting in a mean RACHS score of 4.2 ± 0.3 and mean Aristotle score of 11.1 ± 0.7 for the complete group.

AVSD: atrioventricular septum defect; DORV: double outlet right ventricle; F: female; HLHC: hypoplastic left heart complex; LSVC: left superior vena cava; LV: left ventricle; LVOTO: left ventricular outflow tract obstruction; M: male; MV: mitral valve; PA: pulmonary artery; PS: pulmonary stenosis; RACHS: risk adjustment for congenital heart surgery; RSVC: right superior vena cava; TAPVD: total anomalous pulmonary venous drainage; TGA: transposition of the great arteries; VSD: ventricular septal defect.
mitral valve annulus diameters ‘grew’ to a postoperative z-score of \(+0.7 \pm 0.2\) after surgery and remained at this normalized level after 1-year follow-up (see Fig. 4). These differences were even more striking for patients with HLHC or unbalanced AVSD. Mitral annular size increased in this subgroup from 9.5 ± 0.8 mm preoperatively to 13.9 ± 0.7 mm after LSVC correction (see Fig. 5). The corresponding z-scores were normalized from \(-2.3 \pm 0.2\) to \(0.3 \pm 0.2\) after repair (see Fig. 6). The same effect was present in 4 patients with a large LSVC and a small bridging vein, where we just clipped the LSVC. These patients behaved accordingly but were not included in this study as they did not experience a veno-venous anastomosis as additional repair.

All anastomoses were successfully performed and remained patent in the observed period. After biventricular repair of an unbalanced AVSD, 1 patient developed significant stenosis in the RSVC at the cannulation site below the LSVC–RSVC-anastomosis (see Fig. 7) and had to be balloon-dilated and later stented. The RSVC stayed patent 3 years after surgery and the patient was reballoned twice since then. Permanent anticoagulation treatment with Coumadin® was subsequently necessary.

DISCUSSION

LSVC-related mitral valve inflow obstruction has been described by several different authors in the past [1, 6–8, 10–14, 18, 19]. However, this pathophysiological phenomenon does not occur often and the presence of a large or smaller sized LSVC is generally considered to be of no anatomical relevance, although it seems to have some impact on the growth of left-sided cardiac structures [4, 6]. Liu et al.[2] have reported a subtle but very clear influence of a PLSVC on the structural development of the left heart after reviewing the flow dynamics of 47 foetuses on echo.

This subtle effect is confirmed by some of our elderly patients (>1 year of age) with close to normal-sized mitral valves preoperatively, despite a very large coronary sinus. Neonates and babies below 1 year of age had in contrast very small mitral valves with hypoplastic z-scores below \(-2\) that grew very well after surgical repair.

The anatomical course of all left-sided vena cava through the left atrium in front of the mitral valve annulus has an obstructive potential per se for every patient, but it is rarely considered to be of any haemodynamic significance. Large left-sided vena cava look impressive like a large obstructive hump in front of the mitral orifice on cross-sectional echo studies. Investigators sometimes get the impression that the left atrium is subdivided by the LSVC course [8, 12]. In the presence of an atrial communication, left-to-right shunting is probably increased with consecutive less LV-filling, thus mitral valve and left ventricular hypoplasia might be the consequence [5]. The anatomical location of the LSVC around the mitral annulus might have a purse-string like effect, which may be the reason why mitral valve sizes immediately increased by 1–5 mm in diameter after surgical repair.

However, a normal-sized mitral valve and left ventricle are usually present in most patients despite a moderate or large
LSVC. Different surgical techniques have been reported to solve LSVC-related obstruction [7–9, 11–13, 15, 16, 19] in extreme pathologies. Although most published methods have been able to overcome mitral valve inflow stenosis, only some of these techniques could be considered as anatomical repair [15, 16]. Most of the previously reported methods left the LSVC structure in place, with just less filling by diminishing its size [11–13] or advocated rerouting to the right atrial appendage [9, 19, 20]. A different approach was published by Kreutzer et al. [8], he suggested interposition of a Gore-Tex®-Graft between the 2 vena cava which came close to the idea of an anatomical repair but had to use prosthetic material with the risk of occlusion and no growth potential at all. Following the idea of an anatomical repair as anecdotally published by Reddy et al. and Ugaki et al.[15, 16] on very small patient cohorts, we were able to prove in a larger series that their initially proposed techniques are feasible, safe and effective over the rather short term follow-up of 2.4 years. Reddy has suggested an end-to-side-anastomosis at the level of the right pulmonary artery, passing the mobilized LSVC below the aortic arch. He provided some impressive drawings in his 1997 published ‘how-to-do-it’-paper, but the article, unfortunately, lacks photographs or postoperative angiographies. We were more inspired by the 2010 published article of Ugaki et al., who performed direct end-to-side venous anastomosis anterior and in front of the aortic arch because we think that distortion or compression of the vein is less likely compared to the originally published technique of Reddy. Ugaki applied his technique to 5 patients successfully. He provided no intraoperative pictures either, but at least 1 postoperative angiography demonstrated a patent and adequately sized new surgically created innominate

Figure 3: Four-chamber view before (A) and after (B) anatomical left superior vena cava repair with CS unroofing with significant increase in mitral valve annulus dimensions (a and b).*Left atrial left superior vena cava remnants after unroofing of CS. CS: coronary sinus.

Figure 4: Changes of mitral valve z-scores preoperatively and postoperatively. **P < 0.01.

Figure 5: Changes in mitral valve annular size (in mm) preoperatively and postoperatively for patients with hypoplastic left heart complex, **P < 0.01.

Figure 6: Changes of mitral valve z-scores preoperatively and postoperatively for patients with hypoplastic left heart complex, **P < 0.01.
vein without any obstruction, which is consistent with our results. Despite its low complication and high success rate, the technique has just been applied to a selected subset of patients at our institution as well. The surgically created anastomoses have been checked by echo and cardiac catheter and were all unobstructed and non-occlusive. All veins grew well with no signs of stenosis so far. There was only one intervention (balloon and stent) to an obstructed RSVC, which became narrow below the LSVC–RSVC-anastomosis at the level of the previous cannulation site. This is the only venous complication in the whole cohort, due to technical error, because the very small RSVC was cannulated with an inadequately too large venous cannula. We never considered atrial anastomosis to the right atrial appendage as a real alternative to our approach as some of our patients needed aortic valve operations later, which would have been very challenging technically with the former LSVC crossing anterior and in front of the aortic root.

Complete resection of the inneratrial LSVC course released the posterior mitral annulus and guaranteed unobstructed inflow into the left ventricle. Inneratrial LSVC-resection freed thus at least two-thirds of the mitral annulus, which might have been the reason why larger probes were immediately passing the mitral valve annulus without any surgery to the mitral valve itself. Fortunately, consecutive mitral valve regurgitation due to lesser annular support was not the consequence as the mitral valve chordal apparatus was not surgically damaged. We were glad to see during follow-up that mitral valves and the borderline-sized left ventricles below the affected valves seemed to increase significantly in size according to the consecutive echo studies. We therefore strongly believe that we were able to promote true mitral valve growth by just normalizing the valvar inflow.

CONCLUSION

We report the results of a rather large patient series with HLHC and familiar cardiac malformations after anatomical LSVC repair by direct end-to-side-vein-anastomosis to the RSVC. This simple surgical method reliably relieves mitral inflow obstruction and promotes substantial mitral valve growth for patients with hypoplastic mitral valves and patients with HLHC. The future perspective of this type of anatomical repair should be evaluated on large patient cohorts.

Conflict of interest: none declared.

Author contributions

Robert A. Cesnjevar: Conceptualization; Data curation; Formal analysis; Investigation; Methodology; Project administration; Supervision; Validation; Visualization; Writing—original draft. Frank Harig: Conceptualization; Writing—original draft; Writing—review & editing. Moritz Dietz: Data curation; Formal analysis; Data acquisition. Muhammad Alkassar: Investigation; Methodology; Validation; Visualization. Wolfgang Waellisch: Formal analysis; Investigation; Methodology; Validation; Visualization. André Rueffler: Conceptualization; Formal analysis; Validation; Writing—original draft. Sven Dittrich: Formal analysis; Investigation; Methodology; Supervision; Validation; Visualization; Writing—review & editing. Ariawan Purbojo: Conceptualization; Data curation; Formal analysis; Investigation; Methodology; Supervision; Validation; Visualization; Writing—review & editing.

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