

## Coronary coding in dTGA pre- and post-ASO—verification and necessary corrections following adult CMR

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#### Abstract

| Aims                   | In adult patients with transposition of the great arteries (dTGA) after arterial switch operation (ASO), the coronary artery circulation after neonatal surgical transfer remains a major culprit for long-term sequelae, including myocardial ischaemia and sudden cardiac death. As coronary imaging in paediatric age is often incomplete and classification mainly relies on the surgeon's description in the operation report, we intended to develop a systematic, understandable pattern of the coronary status for each young patient, combining unambiguous coding with non-invasive imaging.   |
|------------------------|--|
| Methods<br>and results | The monocentric prospective study evaluated 89 young adults (mean 23 years) after ASO for dTGA including cardiac mag-<br>netic resonance (CMR) coronary angiography. Following 'The Leiden Convention coronary coding system', we describe the<br>systematic transformation process and provide a graphical illustration considering surgical and imaging views for the six main<br>coronary types, followed by a comparison with adult CMR. Discordance between surgeon's and CMR classification is eval-<br>uated.<br>In seven (7.9%) patients, a discordance between the surgeon's post-operative and the CMR classification was found;<br>therefore, the initial classification had to be corrected according to adult CMR. Three cases (3.4%) with particularly challen-<br>ging coronary variants (intramural and interarterial course, functional common ostium) are presented. |
| Conclusion             | Considering the risks of a possible neonatal coronary misclassification and of increasing additional acquired coronary artery disease with age, reliable cooperation between surgeons, cardiologists, and imaging specialists must be ensured. Therefore, after completion of growth, a systematic pattern of the coronary artery status, combining unambiguous coding with CMR imaging, should be established for each patient.   |

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#### **Graphical Abstract**



Coding transformation process following the Leiden Convention coronary coding system in 89 patients with dTGA after neonatal ASO-verification and

necessary corrections following adult CMR. ASO, arterial switch operation; CMR, cardiac magnetic imaging; dTGA, transposition of the great arteries.

**Keywords** 

dextro-transposition of the great arteries • arterial switch operation • coronary artery classification • Leiden convention coronary coding system • cardiac magnetic resonance tomography

### Introduction

For the last decennia, the neonatal arterial switch operation (ASO) has been used as the surgical procedure of choice to correct the dextrotransposition of the great arteries (dTGA) with intact ventricular septum or with ventricular septal defect. It has shown low mortality and good long-term cardiologic results.<sup>1-3</sup> Currently, long-term survival and freedom from coronary reoperation >25 years after neonatal ASO comprise more than 95% of hospital survivors.<sup>1,2</sup> One of the main challenges during ASO is the transfer of the coronary arteries from the aortic root to the pulmonary artery (PA) root (now neo-aortic root). This can be complicated by the position of the large vessels in relation to one another and by an anomaly of the coronary arteries' origin or branching pattern, which is prevalent in about a third of the cases with a remarkably high variation.<sup>4</sup> From the beginning of the ASO era in the late 1970s, various groups have been working on a standardized description and classification of the various types of coronary anatomies in native dTGA, mainly focusing on the surgical view just before the ASO.<sup>5–10</sup> In 1983, Gittenberger-de Groot, Sauer et al.<sup>6</sup> introduced the so-called 'Leiden Convention', a quite simple but highly applicable classification system for coronary artery types in dTGA. In 2018, Gittenberger-de Groot et al. presented an expanded, broadly applicable system: the so-called 'Modified Leiden Convention'.<sup>11</sup> In 2022, this was followed by the 'The Leiden Convention coronary coding system (LCCCS),' which additionally transfers the surgical view to a universal imaging view.<sup>12</sup> This system now enables a uniform, exact coronary coding by heart surgeons, cardiologists, and imaging specialists.

Within our institution's neonatal ASO programme since 1986, we have established a systematic reassessment programme since 1995, in which patients had undergone standardized cardiological and neurodevelopmental evaluations after neonatal ASO at a mean age of 5,13 10,<sup>14,15</sup> 17,<sup>16</sup> and 23 years,<sup>17,18</sup> in which they had presented with a constantly low late mortality and good cardiological results.

Nevertheless, coronary events are an important factor for increased morbidity and mortality in the long-term, even in previously asymptomatic individuals,<sup>19</sup> and their likelihood increases with specific coronary artery patterns.<sup>20</sup> In this context, a precise description of the coronary anatomy in all patients after ASO for dTGA is important for lifelong follow-up. This information is not systematically available, as it mainly relies on operation reports using different coronary classifications, as well as on often incomplete imaging (e.g. occasional cardiac catheterization) in childhood. The American Heart Association (AHA) guidelines therefore classify the anatomic evaluation of coronary artery patency using coronary catheterization, computed tomography (CT), or cardiac magnetic resonance tomography (CMR) as reasonable in asymptomatic adult patients.<sup>21</sup>

Therefore, the aims of the present study are as follows:

- (1) to evaluate the coronary anatomy in adult patients after ASO by magnetic resonance coronary angiography (MRCA);
- (2) to clarify the use of the LCCCS in a real-life cohort;
- (3) to determine differences between the surgeon's report and the current anatomy that may justify routine coronary artery imaging at least once in adult age in any patient after ASO;

(4) in summary, to establish an exact coronary artery pattern in each patient who had undergone ASO, following the LCCCS, and based on verification of the post-operative surgeon's classification by adult MRCA.

### **Patients and methods**

#### Demographic data

The present monocentric prospective study consisted of 92 unselected young adults examined at a mean age of  $22.8 \pm 2.6$  years (18–29 years, 65% male) who had undergone ASO for dTGA in neonatal age (7 ± 5 days, ) between 1986 and 1998. With respect to coronary artery reinterventions, one patient had undergone a left internal mammary artery (LIMA) bypass to the left anterior descending artery at age 5 years, as described (*Table 3*). Further current investigation results comprising cardiac function, exercise capacity, non-cardiac comorbidity, and quality of life have been recently published.<sup>17,18</sup>

# Surgical management and pre-operative coronary types

During the 12 years' surgical era comprising our study, two experienced paediatric cardiac surgeons were the first surgeons to lead all ASOs in our centre. Both surgeons always followed a common uniform surgical protocol that led to the best possible match of their surgical technique.

From the surgeon's intraoperative perspective, the coronary classification was described before coronary transfer according to the system established in 1983 by Gittenberger-de Groot *et al.*,<sup>6,7</sup> distinguishing six basic types of coronary artery origin and branching patterns (AI, AII, BI, BI, ABI, ABII), including variants with functional common ostium, interarterial, and intramural course (*Table 1* and *Table 3*). The degree of accordance or discordance of the surgeons' classification with the classification from adult CMR was calculated using the four-field table.

ASO was performed as follows<sup>13</sup>:

- Surgical technique based on the Lecompte-modification to establish right ventricle to pulmonary artery continuity.
- Application of a uniform protocol for evaluation of the coronary arteries and careful coronary transfer using the Coronary Button Technique: inspection of the coronary arteries defining the classification type according to Ref. 6. Description of the placement of the large vessels in relation to each other (vertically behind, besides, etc.).
- Transverse transection of the aorta distal to the valve plane.
- First, excision of the right coronary artery from the aorta with a collar and mobilization distally as far as possible (4–5 mm). Then, the same procedure for the left coronary artery.
- Transverse transection of the pulmonary artery immediately before the branch.
- Punching out a circular hole for reimplantation of the left coronary artery into the pulmonary artery base. Suture of the coronary artery continuously. The same procedure is on the right side. The coronary button refers to the portion of the aorta containing the coronary artery ostia.
- Augmentation of the defect in the neo-pulmonary artery with autologous pericardium.

#### Magnetic resonance coronary angiography

A total of 92 patients underwent comprehensive CMR imaging as baseline examination in young adult age. Of these, 89 patients (97%) underwent magnetic MRCA as part of the CMR protocol and met the study's inclusion criteria. Two patients were excluded owing to ventricular arrhythmia, and one because of local artefacts of a mechanical aortic valve prosthesis. The examination was performed on a 1.5 Tesla MR-scanner (Philips Archieva, Best, The Netherlands).

For imaging of the origin and course of the coronary arteries, a free-breathing, navigator-gated, segmented high-resolution 3D steady-state free precession (SSFP) scan with T2 preparation and fat suppression and radial k-space sampling was carried out [resolution:  $1.2 \times 1.2 \times 1.8$  mm with image reconstruction to  $0.7 \times 0.7 \times 0.9$  mm. Parallel imaging (SENSE)-factor: 1.8, TR: 4.2 ms, TE: 2.1 ms, flip angle: 100°]. The diastolic coronary resting period was determined from the cine four-chamber view and the trigger delay was individually adapted. Standard acquisition time was 90 ms per cardiac cycle, with individual reduction as needed. Typical mean scan time was 5 min.

Image interpretation of the coronary origins and course was performed with commercially available software (CVI 42 v5, Circle Cardiovascular Imaging, Calgary, Canada) using multiplanar and curved multiplanar reformation (MPR). For determination of the coronary classification according to the LCCCS,<sup>12</sup> a reformation of the short axis geometry of the neo-artic root at the level of the coronary origins was generated (imaging view). For determination of the course of the coronary arteries, additional MPR and curved MPR were generated as needed. The coronary classification according to the LCCCS was carried out independently by two experienced CMR specialists.

#### Applications of the Leiden Convention coronary coding system

The following systematic applications were used with respect to the LCCCS, which was introduced in 2018<sup>11</sup> and 2022,<sup>12</sup> and its predecessor coding system<sup>6.7</sup>:

- Transformation of the surgeon's coronary type (from the surgeon's written report according to the 'Leiden Convention') into the 'LCCCS' and graphical illustration of the pre-operative 'surgical view' (intraoperative, prior to coronary transfer; *Table 1* and *Figure 1* left).
- Analogue translation of this pre-operative 'surgical view' into the hypothetical pre-operative 'imaging view' with unchanged coding (*Table 1* and *Figure 1* middle-left).
- Adaptation of the hypothetical pre-operative 'imaging view' to the actual late-post-operative 'imaging view' by means of adult MRCA. This demonstrates the adapted coding after ASO, considering the reclassification of the sinuses after surgical coronary artery transfer from the aortic to the neo-aortic root (*Table 1* and *Figure 1* middle-right and right).
- Comparison between the post-operative coding derived from the surgeon's report and the coding of the adult post-operative CMR was carried out in all patients. In case of discrepancy, an analysis and graphical illustration (*Table 2* and *Figure 2*) by comparison of the hypothetical early-post-operative imaging view (derived from the surgeon's report, *Figure 2* middle-left) and the late-post-operative CMR imaging view is provided (*Figure 2* middle-right and right).
- Evaluation and description of special cases of surgical coronary artery transfers during ASO in challenging coronary types and their comparison with the late-post-operative CMR coronary status by means of the LCCCS (*Table 3* and *Figure 3*).
- A general explanation of the LCCCS view orientation (*Table 4*) and the nomenclature used for the coronary artery branches (*Table 5*) is provided.

| Number of  | Number                          |  | Description of coronary   |  |   |
|------------|---------------------------------|--|---|--|---|
| the figure | (%) of<br>patients <sup>a</sup> | Initial surgeon's<br>type (from<br>surgeon's<br>report) <sup>b</sup> | Pre-operative coding<br>(transformed from<br>surgeon's report) <sup>c</sup> | Post-operative coding<br>(considering surgical<br>coronary transfer and sinus<br>reclassification) | arterial origin, course, and<br>branching patterns  |
| 1A         | 64 (71.9)                       | AI   | 1LCx-2R   | 1R–2LCx  | Usual coronary artery type: LCx<br>from left, R from right<br>(posterior) sinus of Valsalva   |
| 1B         | 13 (14,6)                       | AB I   | 1L-2CxR   | 1CxR–2L  | Cx from R: L from left, R and Cx<br>from right (posterior) sinus of<br>Valsalva, Cx with retro-aortal<br>course                               |
| 1C         | 2 (2.3)                         | AII  | 1RLCx   | 2RLCx  | Single left coronary artery: LCx and<br>R from left sinus of Valsalva, R<br>with anterior course  |
| 1D         | 1 (1.1)                         | AB II  | 1RL-2Cx   | 1Cx–2RL  | Inverted R and LCx: L and R from<br>left, R with anterior course; Cx<br>from right (posterior) sinus of<br>Valsalva, with retro-aortal course |
| 1E         | 9 (10.1)                        | BI   | 2LCxR   | 1LCxR  | Single right coronary artery: LCx<br>and R from right (posterior)<br>sinus of Valsalva, LCx with<br>retro-aortal course                       |
| 1F         | 0(0)                            | B II   | 1R–2LCx   | 1LCx–2R  | Inverted LCx and R: LCx from right<br>(posterior) sinus with<br>retro-aortal course, R from left<br>sinus of Valsalva with anterior<br>course |

## Table 1 Systematic transformation process following the Leiden Convention coronary coding system in 89 patients with dTGA after ASO

Representation of the six main anatomical patterns that occur with dTGA. Process from the initial surgeon's coronary artery classification types (from the surgeon's report) to the surgical view, the (hypothetical) pre-operative and the post-operative adult imaging view (from adult CMR, considering sinus reclassification after surgical coronary transfer). Consider *Figure 1*. <sup>a</sup>Number (%) of patients calculated from post-op. imaging view (CMR).

<sup>b</sup>Leiden Convention.<sup>6,7</sup>

<sup>c</sup>The Leiden Convention coronary coding system.<sup>12</sup>

### Results

### Coronary patterns from surgeon's pre-operative report to adult post-operative CMR-systematic transformation process following the LCCCS

The coronary patterns and their frequency distribution are set out in *Table 1*.

Table 1 and Figure 1 describe and illustrate the different steps of the coding transformation process from the surgeon's report to the post-operative CMR for the six main anatomical patterns that occur with dTGA.

- the initial surgeon's coding (from the surgeon's written report) according to the surgical Leiden Convention is translated to the LCCCS (*Figure 1* left).
- (2) the (hypothetical) pre-operative imaging view is formed by analogous translation of the surgical view, coding is unchanged (Figure 1 middle-left).

(3) the post-operative imaging view derived from adult CMR (*Figure 1* right) shows the adapted coding after coronary transfer from the aortic to the neo-aortic root during neonatal ASO, considering sinus reclassification (*Figure 1* middle-right).

#### Comparison between the post-operative surgeon's coding and the adult post-operative CMR coding

Evaluation of the CMR coronary scans showed 100% agreement between the two CMR specialists with respect to the coronary classification following the LCCCS.

Among 89 patients, the post-operative coronary assessments by the surgeon and adult CMR matched in 82 cases. However, discordance was observed in seven patients (7.9%). Specifically, Surgeon 1 performed 38 ASOs (42.7% of the operations), four (4.5%) of which were misclassified according to adult CMR. Surgeon 2 performed 51 ASOs (57.3%), three (3.4%) of which were misclassified according to adult CMR.

Table 2 provides a detailed description of the seven cases, with the initial misclassified surgical and the corrected imaging coding of the

| Table 2 C  | oding correct   | ion following the Leid   | en Convention coronary coding system ar   | nd adult CMR in se   | ven patients  |
|--|---|--|---|--|---|
| Number of  | Number (%)  |  | Coding correction   |  | Rationale and process from initial surgeon's  |
| the figure   | of patients <sup>a</sup>  | Initial misclassified<br>surgeon's type <sup>b</sup> →<br>corrected type <sup>b</sup><br>following adult CMR | Misclassified coding (derived from surgeon's<br>report): pre-op. coding and post-op. coding<br>considering sinus reclassification after<br>coronary transfer <sup>c</sup> | Corrected coding<br>derived from<br>post-op. adult<br>CMR <sup>c</sup> | report to corrected post-op. coding. from adult<br>CMR  |
| 24   | 4 (4.6)   | AI →AB I   | 1LC×-2R → 1R-2LC×   | 1CxR-2L(d)   | In all 4 cases, the adult CMR clearly depicts the retro-aortal course of a small calibre Cx originating from R and a prominent diagonal branch (d) is depicted. This most probably led to the surgeon's misinterpretation of this prominent diagonal branch (d) as Cx, while the actual small calibre Cx was probably not visible or misinterpreted as e.g. an atrial   |
| 2B   | 1 (1.1)   | AB I →A I  | $1L(a)-2CxR \rightarrow 1CxR-2L(a)$   | 1R-2LCx  | branch.<br>In one patient with dextroposition of the heart in situs<br>solitus, the surgeon misinterpreted a small Cx<br>originating from L (sinus 2) as an atrial branch (a) and<br>classified a small side branch from R (originating from<br>sinus 1) as Cx instead. In the adult CMR, a side branch<br>from R is not detected, despite excellent image<br>quality. Instead, the Cx originating from Sinus 2   |
| 3C   | 1 (1.1)   | AB I →B I  | 1L-2CxR→ 1CxR-2L  | 1LC×R-2(rv)  | together with L is clearly depicted.<br>The surgeon misinterpreted an RV branch (rv) as L and<br>transferred it to Sinus 2, leading to a post-operative<br>coding as 1CxR—2L. CMR clearly depicts the distal<br>course of this reimplanted branch towards the free<br>wall of the right ventricle (RV), identifying it as an<br>RV-branch (rv). Instead, L and Cx originate together<br>from Sinus 1 with a proximal retro-aortal course.<br>After the left main bifurcation, L follows the anterior  |
| 5  | 1 (1.1)   | B I → AB I   | 1(c)-2LC×R→1LC×R-2(c)   | 1C-R-2L  | interventricular succis to the LV-apex.<br>Contrary to the aforementioned case, the surgeon<br>misinterpreted L as RV cone branch (c) and<br>reimplanted it in Sinus 2 loco typico, leading to a<br>surgeon's misclassification as $1LCxR-2(c)$ . Adult<br>CMR clearly identifies this relatively small branch as L<br>by depicting its course in the anterior interventricular<br>sulcus towards the LV-apex. The small calibre of L is<br>compensated by an obtuse marginal branch (m) with<br>an intermediate course (originating from Cx) |
| Process from<br><sup>a</sup> Corrected co<br><sup>b</sup> Leiden Conver<br><sup>c</sup> The Leiden C | the initial surgeon'<br>ding: 7.9% of 89 p<br>ntion. <sup>6,7</sup><br>convention coronal | s coronary classification type<br>atients.<br>Y coding system. <sup>12</sup>                                 | to the corrected coding derived from adult CMR, Con:  | isider Figure 2.   |   |

| Table 3 Sy | stematic tra             | nsformation pro  | cess following the Leiden Convention co   | oronary coding system   | in three complex surgical cases   |
|------------|--------------------------|--|---|---|---|
| Number of  | Number (%)               |  | Coding transformation   |   | Rationale and process from initial surgeon's report to  |
| the figure | of patients <sup>a</sup> | Initial pre-op.<br>surgeon's type <sup>b</sup><br>→ Post-op.<br>surgeons's type <sup>b</sup> | Pre-op. surgical coding → post-op. surgical<br>coding (from surgeon's report) <sup>c</sup> considering<br>complex coronary transfer and sinus<br>reclassification | coronary coding from<br>adult CMR, supported by<br>post-op. clinical history <sup>c</sup> | post-op. coding and adult CMR status, considering<br>clinical follow-up   |
| ЗА         | 1 (1.1)                  | A II (double ostium)<br>→ A I  | 1LC×*R*→1R-2LC×   | 1R-2LCx   | The patient had a dorsal ostium from Sinus 1, directly sited at<br>the commissure to Sinus 2, with a functionally common<br>orifice for R and LCx. Taking-off with an acute angle, both<br>followed an interarterial course.<br>Surgeon's procedure comprised a division of R and LCx and a<br>side-by-side reimplantation of R into Sinus 1 and LCx into<br>Sinus 2 of the neo-aorta.<br>Adult CMR confirmed the post-op anatomical status with an<br>acute take-off angle for LCx at 12 to 13 o'clock from Sinus 2<br>and R originating from Sinus 1 without signs of proximal<br>stenosis or kinking.  |
| 8<br>E     | 1 (1.1)                  | BI (double ostium<br>and intramural<br>LCx) → AI   | 2LCx*R → 1R-2LCx  | 1R-2LCx (LCx proximally occluded; LIMA to L)  | Up to now, the patient has shown a bland clinical follow-up<br>The patient had a functionally common ostium from Sinus 2,<br>sited nearby the commissure to Sinus 1.<br>The surgeon's procedure first comprised a division of R and<br>LCx. After mobilization, R could be reimplanted without<br>obstruction into Sinus 1. However, LCx showed an<br>acute-angle take-off and a proximal intramural and<br>interarterial course before leaving the aortic wall. After<br>mobilization and longitudinal splitting of the narrow vessel,<br>LCX occluded around the neo-aortic reimplantation site in the<br>longer term, leading to electrocardiogram repolarization<br>disorders under stress. At age 5 years, the patient underwent<br>a successful LIMA to L bypass-grafting, then showed a bland<br>clinical course again.<br>Adult CMR confirmed the anatomical post-bypass situation,<br>already documented in a former invasive coronary |
|            |                          |  |   |   | angiography   |

6

Continued

| Number of  | Number (%)   |  | Coding transformation   |   | Rationale and process from initial surgeon's report to   |
|--|--|--|---|---|--|
| the figure   | of patients <sup>a</sup>   | Initial pre-op.<br>surgeon's type <sup>b</sup><br>→ Post-op.<br>surgeons's type <sup>b</sup> | Pre-op. surgical coding → post-op. surgical<br>coding (from surgeon's report) <sup>c</sup> considering<br>complex coronary transfer and sinus<br>reclassification | coronary coding from<br>adult CMR, supported by<br>post-op. clinical history <sup>c</sup> | post-op. coding and adult CMR status, considering<br>clinical follow-up  |
| g  | 1 (1.1)  | ABI (intramural L) →<br>ABI  | 11*-2CxR → 1CxR-2L  | 1CxR-2L (L proximally<br>occluded and collateralized<br>via R)                            | L originated from Sinus 1 and presented an acute-angle take-off<br>and a proximal intramural and interarterial course. R took off<br>from Sinus 2 and Cx originated from the proximal R and had a<br>posterior looping course, as usual in this coronary artery type.<br>The surgeon's procedure consisted of a difficult mobilization<br>and excision of both coronary arteries and a complicated<br>reimplantation of RCx into Sinus 1 and L into Sinus 2.<br>L occluded in the area of the reimplantation site in the longer<br>term, documented by invasive angiography. Sufficient natural<br>retrograde collateralization via R and Cx to L had developed,<br>so that bypass surgery was dispensed so far.<br>Adult CMR confirmed the described anatomical situation with a<br>proximal occlusion of L. Up to now, the 20-year-old patient<br>has shown a bland clinical follow-up |
| Process from the<br><sup>a</sup> Complex cases 3<br><sup>b</sup> Leiden Convention<br><sup>c</sup> The Leiden Conv | nitial pre-op. to the<br>4% of 89 patients<br>6.7<br>ention coronary cod | oost-op. surgeon's classifical<br>ng system. <sup>12</sup>                                   | tion, followed by the coding derived from the adult CMR, consi  | idering post-op. clinical history. Con  | sider Figure 3.  |

coronaries, as well as the most probable rationale for the surgeon's misclassification. Figure 2 illustrates the process of coding correction, showing the misclassified pre-operative surgical coding (Figure 2 left) and the misclassified early-post-operative coding according to the surgeon's report (Figure 2 middle-left). The latter is provided in the (hypothetical) imaging view orientation for better comparison with the actual post-op adult imaging view (Figure 2 middle right), which is derived from the CMR (Figure 2 right). A comparison between the misclassified earlypost-operative (Figure 2 middle-left) and the corrected late-post-operative (Figure 2 middle-right) imaging view illustrates the rationale for the surgeon's misclassification and the need for coding correction following the adult CMR. For a better understanding, the initially misinterpreted coronary branches and relevant side branches depicted in the CMR have been inserted into the code formula with a lowercase letter in brackets (atrial (a), cone (c), diagonal (d), obtuse marginal (m), and right ventricular (rv) branch).

#### Long-term fate of high-risk coronary patterns from initial surgeon's procedure to actual CMR status

Three of 89 patients (3.4%) had a complex coronary artery variant representing a particularly challenging pattern for the surgeon and, in the longer term, a possibly increased risk of severe coronary complications and sudden cardiac death, respectively. These show a proximal interarterial course (indicated by an asterisk, \*), in two cases paired with a double (functional common) ostium for the right and left coronary arteries, and in two cases associated with a proximal intramural course of one or more coronary arteries.

Table 3 provides details to each patient and Figure 3 illustrates the different steps of the coding transformation process: (1) the LCCCS surgical view and coding, following the surgeon's written report (Figure 3 left); (2) the analogue translation to the hypothetical pre-operative imaging view and coding (Figure 3 middle-left); (3) the post-operative imaging view and adapted coding taking into account sinus reclassification as well as individual coronary transfer, showing concordance of the surgeon's report with the adult CMR (Figure 3 middle-right and right). In all three patients, the great vessels were in d-transposition providing a vertical anterior–posterior axis, in one case the aorta even had a slight left position to the pulmonary artery.

#### Discussion

The steadily growing group of adult patients after ASO for dTGA directs an important focus of interest to the individual anatomical and functional patterns of the coronary arteries after surgical coronary transfer into the neo-aorta in neonatal life.

#### Systematic transformation process of coronary artery patterns following the LCCCS

To our knowledge, the present study provides for the first time a step-by-step description and graphical presentation of the systematic coding transformation process to and within the Leiden Convention coronary coding system (LCCCS), starting from the surgeon's report to the post-operative status and supported by a comparison with adult CMR imaging. Based on the surgeon's historical pre-operative coronary classification (according to Refs. 6,7), the individual coronary pattern now considers the post-operative reclassification of the neo-aortic sinuses after coronary transfer, which is obligatory in the LCCCS,<sup>11,12</sup> as well as surgical peculiarities in coronary transfer.

It should be considered that information about the exact course of the coronary arteries and thus the coronary classification is not available in most adolescent patients with an uncomplicated course after neonatal ASO. Notably, routine cardiac catheterization without selective coronary angiography, as often carried out in infancy, primarily serves to visualize and secure a stenosis-free exit of the reimplanted coronary arteries as well as a kink-free course of the neo-pulmonary artery.

## Coding correction of misclassified surgeon's coronary artery patterns

In our study, we find a discordance between the coronary classification from the surgeon's report and from adult CMR imaging in 7.9% of the patients. As MRCA provides an excellent depiction of the coronary anatomy,<sup>22,23</sup> the initial surgeon's coding must be interpreted as misclassified and therefore must be corrected. This misclassification by the surgeon most probably is favoured by the limited field of vision during neonatal ASO and the small calibre of the coronary branches in neonatal age. By this, the main course of the coronary branches is generally not visible during ASO, whereas it is clearly depicted by MRCA.

The correction of the coronary classification is of clinical relevance, as it may lead to a change in the patient's coronary artery-related risk estimation.<sup>1</sup> Therefore, this information may influence the patient's prognosis and the strategy of clinical follow-up.

With increasing age, the exact coronary anatomy may become important in the context of acquired, atherosclerotic coronary artery disease. In acute coronary syndromes, unusual coronary anatomy is a special challenge during urgent coronary catheterization,<sup>24,25</sup> and incorrect information about the coronary pattern may lead to wrong therapeutic decisions and worse outcomes. In this context, prominent side branches of the main coronary arteries should be considered clinically relevant for the myocardial blood supply. Therefore, we suggest adding relevant coronary side branches to the LCCCS by using lower case letters in brackets [atrial (a), cone (c), diagonal (d), obtuse marginal (m), and right ventricular (rv) branch].

# Long-term follow-up of high-risk coronary artery patterns

Three of 89 patients had pre-operative complex coronary patterns. After primarily successful surgical coronary transfer, a permanent increased risk of coronary events or even sudden cardiac death has been the subject of controversial discussion.<sup>1,7,9,26–29</sup> In our study, the two cases with proximal intramural course showed ostial occlusion of these reimplanted vessels in adult CMR. Those patients must be considered at risk for future ischaemic coronary events. Therefore, the need for regular cardiac monitoring remains, especially as dTGA patients may experience less or no ischaemia-induced cardiac symptoms because of partial myocardial denervation after ASO.<sup>30–32</sup>

# Application of CMR for coronary artery imaging

According to AHA guidelines, a baseline examination to determine the origin and proximal course of the coronary arteries is reasonable to perform in young adults with dTGA after ASO,<sup>21</sup> whereas the European Society of Cardiology (ESC) guidelines question the necessity of routine coronary imaging.<sup>33</sup> Our study reveals a remarkable incidence of coronary misclassifications up to adult age and therefore supports the strategy of routine baseline non-invasive coronary imaging.

MRCA provides reliable imaging of coronary artery ostia and proximal segments, comparable to CT. It offers additional information about the exact course of the individual coronary artery branches including potentially malignant features that may have evolved during growth.<sup>22,23</sup> While MRCA, other than CT, cannot visualize coronary artery calcifications and has limited imaging quality for the



**Figure 1** Graphical representation and adult CMR of coronary status in dTGA. Coding transformation process in 89 patients following the Leiden Convention coronary coding system (LCCCS) and adult CMR for the 6 main anatomical patterns (A to F) occurring with dTGA. Consider Table 1, 4 and 5. Left: 'pre-operative surgical view' derived from surgeon's report. The Leiden code for the different coronary patterns is depicted in Table 1. Middle-left: hypothetical 'pre-operative imaging view', generated from the surgical view by analogue translation. The Leiden code keeps unchanged. Middle-right: 'post-operative imaging view', graphical image derived from adult CMR. The Leiden code is changed according to a sinus reclassification after surgical coronary transfer from the native aorta (now Neo-PA) to the neo-aorta. *Right:* (post-operative) adult CMR: multiplanar reformation (MPR) in the 'imaging view' orientation at the level of the coronary artery origins.



**Figure 2** Graphical representation and adult CMR image of coronary coding correction. Coding transformation process in seven patients (A: four patients; *B*, *C*, *D*: each one patient) from the surgeon's coronary classification to coding correction after adult CMR, following the LCCCS. Consider *Table 2*, *4*, and *5*. *Left:* 'pre-operative surgical view' derived from the surgeon's report. Consider *Table 2* for Leiden code. *Middle-left:* hypothetical early 'post-operative imaging view', derived from the surgeon's report. The Leiden code is changed according to sinus reclassification after coronary transfer from native Ao to Neo-Ao. *Middle-right:* late 'post-operative imaging view', graphical image derived from adult CMR with corrected coding compared with initial surgeon's report. Discrepancy between hypothetical early-post-operative and late CMR-derived Leiden code demonstrates the surgeon's misinterpretation (compare middle-left and middle-right). *Right:* adult CMR: multiplanar reformation (MPR) at level of coronary artery origins and (curved) MPR of distal coronary course. *Dashed, faded arteries:* probably not visible to the surgeon during neo-natal ASO.



**Figure 3** Graphical representation and adult CMR image of complex surgical cases. Coding transformation process in three patients with dTGA after ASO with special surgical challenges (A, B, C) following the LCCCS and adult CMR, considering post-operative clinical history. Consider *Table 3, 4, and 5*. *Left:* 'pre-operative surgical view' following surgeon's report, demonstrating pre-operative complex coronary status with double coronary ostium (A, B), and interarterial (A, B, C) and intramural course (B, C). Consider *Table 3*. *Middle-left:* 'pre-operative imaging view', generated from the surgical view by analogue translation. The Leiden code keeps unchanged. *Middle-right:* 'post-operative imaging view', graphical image generated from adult CMR, supported by the surgeon's report. The Leiden code is changed owing to sinus reclassification after individual complex coronary transfer from the native Ao (now Neo-PA) to the Neo-Ao and patient history. *Right:* adult CMR: multiplanar reformation (MPR) at the level of coronary artery origins and (curved) MPR of distal coronary and bypass course. *LIMA*: left internal mammary artery.

very distal coronary artery branches, it is free of ionizing radiation. Moreover, CMR uniquely offers detailed information about cardiac function, viability, great vessel geometry, and pulmonary perfusion in one examination. Therefore, regular CMR follow-up examinations are recommended in patients with dTGA after ASO to assess neo-aortic size, branch pulmonary arteries, and ventricular and valvular functions.<sup>21,34</sup> Given our patients' average age of under 25 years, where acquired coronary artery calcification is unlikely and the

#### Table 4 Surgical and imaging view orientation within the Leiden Convention coronary coding system

| View          | Explanation of the orientation                                   |
|---------------|--|
| Surgical view | Cranial view from the non-facing sinus (NF) of the               |
| orientation   | aortic valve (Ao).   |
|               | Looking inwards (blue arrow with inscription                     |
|               | 'look') towards the posterior positioned                         |
|               | pulmonary artery (PA):   |
|               | <ul> <li>Right hand is Sinus 1, counterclockwise</li> </ul>      |
|               | followed by Sinus 2  |
|               | <ul> <li>Coronary arteries are enumerated</li> </ul>             |
|               | counterclockwise (blue dashed arrow with                         |
|               | inscription ' <b>turn</b> ')                                     |
| Imaging view  | Caudal view directed upwards from the left                       |
| orientation   | ventricle  |
|               | Looking outwards (blue arrow with inscription                    |
|               | ' <b>look</b> ') from the NF of the Ao:                          |
|               | <ul> <li>Right hand is Sinus 1, clockwise followed by</li> </ul> |
|               | Sinus 2  |
|               | <ul> <li>The coronary arteries are enumerated</li> </ul>         |
|               | clockwise (blue dashed arrow with inscription                    |
|               | 'turn')  |

# Table 5 Nomenclature of the coronary arteries within the Leiden Convention coronary coding system

| LCx | Left coronary artery                           |
|-----|--|
| L   | Left anterior descending coronary artery (LAD) |
| Cx  | Left circumflex artery                         |
| R   | Right coronary artery (RCA)                    |
| а   | Atrial branch                                  |
| с   | Cone branch                                    |
| d   | Diagonal branch                                |
| m   | Obtuse marginal branch                         |
| rv  | Right ventricular branch                       |
| *   | Symbol for interarterial course                |

avoidance of ionizing radiation is preferred, we opted for a comprehensive CMR examination including MRCA over CT for visualization of the coronary arteries.  $^{34}$ 

#### **Study limitations**

The findings of our study, carried out on a uniform group of patients with dTGA, were based on precise surgical operation reports from two experienced surgeons who consistently followed the early Leiden Convention.<sup>6,7</sup> However, the single-centre design of our study limits the generalization of our results regarding the incidence of discrepancies between the surgeon's assessments and those derived from adult CMR. Notably, in the case of less conclusive surgical reports, this transformation process may not be so straightforward. In those

cases, the value of non-invasive imaging to visualize coronary anatomy and establish the exact coronary coding is even higher. Contraindications as certain medical implants may prohibit CMR, and the value of MRCA may be limited in patients with relevant arrhythmia or local artefacts due to mechanical aortic valve prosthesis, as present in three patients in our cohort. In those cases, CT coronary angiography is an alternative to MRCA, accepting the application of ionizing radiation in young patients.

## Conclusions

After completion of growth, a systematic anatomic pattern of the coronary arteries, including a radiation-free, non-invasive CMR imaging, should be established for each patient after neonatal ASO for dTGA. Since the risk of additional acquired coronary artery disease increases with age, a safe and reliable cooperation and understanding between imaging specialists, cardiologists, and cardiac surgeons needs to be ensured. For this concept, the Leiden Convention coronary coding system provides an optimal basis.

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**Conflict of interests:** Any conflicts of interest have been disclosed for all authors

## Consent

The study was approved by the local Ethical Medical Committee (Medical Faculty RWTH Aachen University, Aachen, Germany, Nr. EK 243/14), and written informed consent was obtained from each patient. The funding agency was not involved in data interpretation.

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### Data availability

The data underlying this paper cannot be shared publicly owing to data protection regulations. The data will be shared on reasonable request to the corresponding author.

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