Venovenous malformation: a common finding after Kawashima operation

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Abstract

Objectives: It has been reported that systemic venovenous malformation (VVM) can develop in patients with interrupted inferior vena cava (IVC) and univentricular type of congenital heart disease who undergo superior vena cava to pulmonary artery connection (Kawashima operation). These malformations can lead to profound systemic desaturation postoperatively. However, there have been few reports that characterise the prevalence, anatomic details and clinical correlations of these systemic VVM arising after Kawashima operation. In this study, we describe our experience with VVM after Kawashima operation, and discuss issues regarding their evaluation and postoperative management.

Methods: Eight patients with median age 19 months (range: 5—238) who underwent Kawashima operation were subjected to postoperative angiography, prospectively. Sites of VVM origin and entry, as well as their course, were documented. The presence of pulmonary arteriovenous malformations (AVMs) was also documented. Results: At median follow-up of 31 months (range: 16—72 months), a total of 14 VVM were found in different supra- and infra-diaphragmatic sites in six patients (75%); four of them had concomitant pulmonary AVM while the remaining two patients had only pulmonary AVM. Conclusions: Our findings suggest that systemic VVM can occur frequently after Kawashima operation and can produce significant desaturation postoperatively, and hence we support hepatic incorporation. Performing detailed angiographic studies of the supra- and infra-diaphragmatic systemic veins in routine assessment of patients before Kawashima operation is, probably, warranted.

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Keywords: Kawashima operation; Venovenous malformation; Pulmonary arteriovenous malformation; Interrupted inferior vena cava; Cavopulmonary connection

1. Introduction

Venovenous malformation (VVM) causing significant hypoxaemia may develop after Kawashima operation in patients with single ventricle anomaly and interruption of inferior vena cava (IVC), where lower extremity venous return to superior vena cava (SVC) is maintained through azygous vein, hemi-azygous vein, or both [1—4]. The prevalence of such malformation is considered as a common finding after cavopulmonary (Glenn and Fontan) operation and varies from 20% to 31% [5—7]. However, this matter has not received great deal of attention in the literature with regard to Kawashima operation.

We encountered a patient who had severe desaturation after Kawashima operation. The cause of desaturation was not determined in spite of thorough investigations, and pulmonary arteriovenous malformation (AVM) was excluded. In an effort to exhaust all possible causes, a venogram was performed along the azygous vein and two large VVM were detected shunting blood from the azygous vein into the hepatic venous system and were responsible for the significant desaturation. This experience led us to review all our patients with interrupted IVC who underwent Kawashima operation at our hospital.

In this study, we report eight patients who underwent Kawashima operation with special emphasis on the detection and description of VVM, relevant haemodynamic and echocardiographic data, and the relation to postoperative desaturation. The presence of pulmonary AVM was also documented.

2. Methods

Between April 2002 and March 2008, 12 patients had undergone Kawashima operation at our hospital. Four patients were excluded from the study; three of them could not be traced (living outside Saudi Arabia), and one patient with acceptable oxygen saturation, his guardian, refused to participate in the study. The remaining eight patients were prospectively studied for the presence of VVM after obtaining informed consent from their parents/guardians.
2.1. Data collection

The study was approved by the Institutional Review Board of King Faisal Specialist Hospital and Research Center, Jeddah, Saudi Arabia. Medical charts, echocardiography, cardiac catheterisation, and operative data were reviewed, retrospectively. Prospectively, at the time patients were enrolled in the study, they underwent echocardiography, cardiac catheterisation, and lung perfusion scan to evaluate the presence of VVM and pulmonary AVM.

Data were divided into three different phases:

(A) Pre-Kawashima data: It included demographic data (age, weight and haemoglobin level), echocardiographic data (cardiac diagnosis, function of the systemic ventricle and the function of the systemic atrioventricular valve), and cardiac catheterisation data (pulmonary artery pressure (PAP) in mm Hg, pulmonary vascular resistance (PVR) in wood’s unit (WU) and the end-diastolic pressure of the systemic ventricle (EDP) in mm Hg).

(B) Peri-Kawashima data: This included operative data and associated procedure, intensive care unit stay, hospital stay and room-air percutaneous oxygen saturation (RA SpO₂) at the time of discharge from the hospital. RA SpO₂ was obtained at rest for all patients using commercially available pulse oximeter.

(C) Post-Kawashima data: This comprised a prospective review of clinical history, RA SpO₂, weight, haemoglobin level, chest X-ray, echocardiography, cardiac catheterisation and nuclear lung perfusion scan. RA SpO₂ during last follow-up in the clinic was obtained at rest for all patients. Echocardiographic assessment includes ventricular function, and the function of systemic atrioventricular valve. Cardiac catheterisation data included systemic ventricular EDP, PAP, PVR and the presence of cavopulmonary gradient. During the procedure, agitated saline injection in the pulmonary artery in close proximity to the injected contrast in the proximal pulmonary arterial (PA) angiography, (2) echocardiographic detection of bubbles in the pulmonary atrium after direct injection of agitated saline in the pulmonary artery and/or (3) positive extra-pulmonary isotope uptake by brain or kidney during the lung perfusion scan after isotopic injection in a peripheral vein in the upper extremities.

Systemic VVM was defined as a venous channel allowing blood flow away from the pulmonary circulation to the hepatic circulation, atrium or pulmonary veins. VVM, when present, were characterised according to the size, site and destination. The widest diameter VVM, left SVC and right SVC were measured. The calibre of VVM was graded relative to the diameter of the largest SVC; <25% was graded as small, between 25% and 50% were graded as medium, and >50% were graded as large.

2.3. Statistical analysis

Statistical analysis was performed with SPSS statistical program (SPSS 12 Inc., Chicago, IL, USA). Data were presented as mean ± standard deviation or median with ranges or percentages, as appropriate. The Shapiro—Wilk normality test was used to assess normal distribution. Differences in categorical variables were analysed by means of the χ² analysis, and differences in continuous variables were analysed by Student’s t-tests. The small number of our group did not allow for regression analysis. Differences were considered to be statistically significant when p value was less than 0.05.

3. Results

3.1. Pre-Kawashima data (Table 1)

The median age at the time of Kawashima operation was 19 months (range: 5–238 months) and the median weight was 8 kg (range: 5.6—47.5 kg). Five patients (62.5%) had a systemic ventricle of right ventricular morphology. Haemoglobin level was ranging between 143 and 183 g l⁻¹ (median; 160 g l⁻¹). Three patients had abdominal situs inversus and two patients had abdominal situs ambiguous. Four patients had bilateral SVC, three patients had left SVC and one patient had right SVC. None of the patients with bilateral SVC had a bridging innominate vein. Two patients had azygous continuation of the IVC, three patients had hemi-azygous continuation and three patients had both azygous and hemi-azygous continuation. Echocardiography showed no significant AVV regurgitation, and all patients had good systemic ventricular function, except one patient, who had fair-to-good function. Cardiac catheterisation documented mean PAP of 15.8 ± 3 mm Hg, mean EDP of 11.1 ± 4 mm Hg and PVR of 1.6 ± 0.1 WU.

3.2. Peri-Kawashima data

The cavopulmonary anastomosis was performed on cardiopulmonary bypass through midline sternotomy. An end-to-side anastomosis of the SVC to the PA was con-
Table 1. Patients demographic, echocardiographic, haemoglobin, saturation and haemodynamic data; before and after Kawashima operation.

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (mo)</th>
<th>Wt (kg)</th>
<th>Diagnosis</th>
<th>SVC site</th>
<th>Azyg/hemi cont</th>
<th>Hb (g l⁻¹) ECGH</th>
<th>Pre-Kawashima data</th>
<th>Post-Kawashima data</th>
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- **Pre-Kawashima data:**
  - **Hb (g l⁻¹) ECGH:** The range is from 94% to 100%, with a mean of 98%.
  - **AVR:** The range is from 0.1 to 0.9 WU, with a mean of 0.5 WU.
  - **ECHO Catheterisation data:** The range is from 120 to 200 g l⁻¹, with a mean of 154 g l⁻¹.

- **Post-Kawashima data:**
  - **Hb (g l⁻¹) ECGH:** The range is from 90% to 100%, with a mean of 96%.
  - **AVR:** The range is from 0.1 to 0.9 WU, with a mean of 0.5 WU.
  - **ECHO Catheterisation data:** The range is from 120 to 200 g l⁻¹, with a mean of 154 g l⁻¹.

**3.3. Post-Kawashima data**

Echocardiographic, RA SpO₂, haemoglobin level and catheterisation data are shown in Table 1. RA SpO₂ range was 70–88% with a mean of 78%. Haemoglobin level was ranging between 120 and 200 g l⁻¹ (median; 154 g l⁻¹). Cardiac catheterisation was performed between 16 and 72 months after Kawashima operation (median 31.5 months). No significant pressure gradient was found between the SVC and the PA, and no pulmonary venous obstruction could be detected in all patients. The mean PAP was 14 ± 3 mm Hg, mean EDP was 9 ± 3 mm Hg and PVR was 0.9 ± 0.6 WU.

Six patients, out of eight (75%), had clinically significant desaturation of more than 5% from immediate post-Kawashima saturation compared to saturation at last follow-up. All patients with desaturation had either VVM or AVM or both. Two patients (cases #1 and #2) did not have clinically significant desaturation (<5%), despite in case #1, there was AVM and a small VVM, and in case #2, there was a small VVM and another large one to the hepatic circulation.

Total of 14 measurable VVM were detected in six out of eight patients (75%). Five were of large, four were of medium and five were of small calibres. Eleven out of 14 VVM were detected infra-diaphragmatically, and three patients had both supra- and infra-diaphragmatic VVM (Table 2). The eventual destinations of these VVM were from the azygous and hemi-azygous systems to the hepatic venous system, portal system, pelvic, paravertebral plexus, perirenal system and right upper pulmonary vein. Examples are shown in Figs. 1 and 2.

In one patient (case #3), coil embolisation was performed for a small VVM routing to right upper pulmonary vein with no increase in room-air oxygen saturation. However, the two patients (cases #4 and #6) who developed pulmonary AVM only required dilatation and stenting of the pulmonary artery banding site with ensuing increase in RA SpO₂.

Two patients (cases #5 and #8) underwent hepatic venous incorporation. One of them had improvement of RA SpO₂ from 70% to 90% postoperatively. However, the other one died intra-operatively from accidental aortic tear.

In our analysis, pre-Kawashima mean EDP was significantly higher among the six patients with positive VVM.
(12.5 ± 3.7 mm Hg) compared to the two patients without VVM (7 ± 0.0 mm Hg) with a p value of 0.015. However, mean PAP (16 ± 4 mm Hg vs 14.5 ± 0.7 mm Hg, p = 0.522) was not significantly different. The difference between PAP and EDP (trans-pulmonary gradient) was analysed, and it was not significant (p = 0.539).

From the post-Kawashima haemodynamic data, the mean PAP and mean PVR were significantly higher among the six patients with positive VVM compared to the two patients without VVM; 15.8 ± 2.5 mm Hg versus 9.5 ± 0.7 mm Hg (p = 0.001), and 1.1 ± 0.5 WU versus 0.4 ± 0.02 WU (p = 0.018), respectively. However, mean EDP (10.5 ± 3.7 mm Hg vs 8 ± 1.4 mm Hg, p = 0.225) was not significantly different, and the difference between PAP and EDP (trans-pulmonary gradient) was not significant too (p = 0.152).

There was no significant correlation between VVM and other variables such as site of SVC, haemoglobin level and RA SpO2.

Contrast echocardiography and nuclear lung scan documented the presence of pulmonary AVM in six cases. However, three of them were confirmed with the proximal pulmonary artery angiography. Statistical analysis showed no significant association between the incidence of AVM and either preoperative or postoperative haemodynamic vari-

<table>
<thead>
<tr>
<th>Case no.</th>
<th>VVM no.</th>
<th>VVM</th>
<th>AVM</th>
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<tr>
<td></td>
<td>Size</td>
<td>Site</td>
<td>Destination</td>
</tr>
<tr>
<td>1</td>
<td>Small</td>
<td>Infra-diaphragmatic</td>
<td>Pelvic venous plexus</td>
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<td>2</td>
<td>Small</td>
<td>Supra-diaphragmatic</td>
<td>Paravertebral plexus</td>
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<td>Large</td>
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<td>Hepatic</td>
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<td>3</td>
<td>Small</td>
<td>Supra-diaphragmatic</td>
<td>Right upper pulmonary vein</td>
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<td>Medium</td>
<td>Infra-diaphragmatic</td>
<td>Perirenal venous plexus</td>
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<td>Medium</td>
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<td>5</td>
<td>Small</td>
<td>Supra-diaphragmatic</td>
<td>Internal mammary vein</td>
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<td>Large</td>
<td>Infra-diaphragmatic</td>
<td>Hepatic vein</td>
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<td>Medium</td>
<td>Infra-diaphragmatic</td>
<td>Splenic vein</td>
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<td>6</td>
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<tr>
<td>7</td>
<td>Small</td>
<td>Infra-diaphragmatic</td>
<td>Pelvic plexus</td>
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<td>Large</td>
<td>Infra-diaphragmatic</td>
<td>Perirenal plexus</td>
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<td></td>
<td>Medium</td>
<td>Infra-diaphragmatic</td>
<td>Hepatic vein</td>
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<tr>
<td>8</td>
<td>Large</td>
<td>Infra-diaphragmatic</td>
<td>Hepatic vein</td>
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<tr>
<td></td>
<td>Large</td>
<td>Infra-diaphragmatic</td>
<td>Paravertebral plexus</td>
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ables such as EDP, PAP, PVR, single ventricle diagnosis, site of SVC, haemoglobin level and RA SpO2.

4. Discussion

With meticulous angiography above and below the diaphragm, systemic VVM can be found frequently after Kawashima operation (75%). This is similar to that reported by Stumper and colleagues [1] 80% and Kaneko and colleagues [3] 60%. This high frequency is because the presence of VVM was actively sought. Failure to visualise VVM before surgery occurs because low inferior (or modified) venography is not performed as a part of routine diagnostic evaluation. Inferior venography should be performed in all patients as most of VVM (11 out of 14) observed in our study were infra-diaphragmatic.

It appears from our data and other published reports [1—4] that VVM can manifest at any time (16–72 months) after Kawashima operation and can be of varying sizes, numbers and can lead to clinically significant desaturation. This desaturation was experienced in four out of six VVM patients; two of them required hepatic venous incorporation.

There are multiple factors that contribute to the development of venous collateral channels after Kawashima operation. One factor may be related to the patient’s native venous anatomy [5]. Another factor is the pressure differential between the SVC and pulmonary atrium [7].

The embryological development of IVC may help to explain the cause of VVM. During the course of IVC development, many venous channels are known to disappear [8]. We agree with others [5,7] that such channels re-appear when the cavae are subjected to increased pressures after cavopulmonary anastomosis. We believe that these VVM channels may be present before surgery, yet remain undiagnosed until the circulation is re-routed with production of decompression channels away from the pulmonary circulation. However, it is not clear why some patients develop VVM channels after Kawashima operation and others do not, and why the extent of channel development varies so greatly.

It seems possible that VVM channel development may correlate with the pressure differential between the bidirectional Glenn circuit presented by the PAP and the systemic ventricle EDP. Magee and colleagues [7] reported that the gradient between the SVC and the pulmonary atrium was independently associated with the presence of VVM. As all our patients did not have obstruction at the anastomotic site, a connection between SVC and the azygous system may allow for decompression of the SVC venous blood away from the pulmonary circulation. However, our data did not show any significant association between VVM and the trans-pulmonary gradient.

The most significant potential complication associated with VVM is systemic desaturation. Whether VVM develops early or late, they should always be considered as a potential cause when patients with Kawashima physiology develop unexplained hypoxia or desaturation. The awareness of the presence of VVM provides the opportunity for timely and precise intervention if symptomatic cyanosis occurs, without the need for exhaustive investigation of the patients in the early period after surgery. We agree with others [2—4,9] that earlier referral for hepatic vein redirection in the patient with good haemodynamics for conversion to a total cavopulmonary anastomosis may be considered.

Pulmonary AVMs are also reported to occur in 21–58% of patients after Kawashima operation [10—12]. Their incidence in our study was very high (75%), and was associated with VVM in four of our patients. Their cause remains unknown; however, mal-distribution and non-pulsatile pulmonary blood flow [13] and lack of hepatic factor [10] due to diversion of normal hepatic venous flow from the pulmonary circulation have been implicated in their pathogenesis. This is very important in patients with interrupted IVC when the cavopulmonary anastomosis is considered to be the definitive procedure. Although there is no clear explanation for the association between AVM and VVM, but they share their clinical effect on oxygen saturation and surgical management with hepatic venous incorporation.

In summary, clinically important VVM and pulmonary AVM may form in the majority of patients after Kawashima operation. The diagnosis of pulmonary AVM after Kawashima operation should not preclude investigating the presence of VVM as they may exist in the same patient as four out of eight of our patients had both pulmonary AVM and VVM. Awareness of VVM may guide the surgical plan of patients with interrupted IVC.

5. Conclusions

Systemic VVM can occur frequently after Kawashima operation and can produce clinically significant desaturation during follow-up. Performing detailed angiographic studies through the femoral vein and azygous or hemi-azygous system to rule out pre-Kawashima VVM, especially below diaphragm, which is not included in routine diagnostic cardiac catheterisation may be warranted. Moreover, although it was not shown in our study, early referral of Kawashima patients to complete the Fontan circuit by incorporating the hepatic vein may decrease the development of both VVM and pulmonary AVM.

References


