Innominate artery compression of the trachea in patients with neurological or neuromuscular disorders

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Abstract

OBJECTIVES: The purpose of this study was to review and assess our surgical management of innominate artery compression of the trachea (IACT) in patients with neurological or neuromuscular disorders (NMDs).

METHODS: Thirty patients with NMD who underwent surgical treatment for IACT at Kobe Children's Hospital and Kobe University Hospital from 2002 to 2012, were enrolled in this retrospective study. The clinical outcomes of preventive elective surgery for IACT (Group A, n = 20) were assessed and compared with those of emergent surgery (Group B, n = 10).

RESULTS: A total of 27 patients underwent innominate artery transection (17 in Group A and 10 in Group B), and 3 patients in Group A underwent innominate artery reimplantation using prosthetic graft interposition. No operative or early death occurred. There were no cases of postoperative mediastinitis or neurological complications. The operative benefits in Group A included a smaller skin incision, more limited sternotomy, less blood loss, shorter operative time and shorter hospital stay, compared with Group B. No blood transfusion was required in Group A. The number of patients in whom cerebral circulation was assessed before surgery in Group A was significantly higher than those in Group B.

CONCLUSIONS: Preventive elective surgery for IACT provides many advantages, including minimally invasive procedures and successful postoperative outcomes without neurological complications in patients with NMD. Because this surgical management can prevent the tragic occurrence of a tracheo-innominate artery fistula or an exacerbation of tracheomalacia, it would be an optimal surgical treatment for IACT to improve the quality of life in patients with NMD.

Keywords: Innominate artery compression of the trachea • Neurological or neuromuscular disorders • Tracheoinnominate artery fistula • Tracheomalacia • Preventive elective surgery

INTRODUCTION

Recent advances in respiratory support technologies have had a significant impact on the natural history of patients with neurological or neuromuscular disorders (NMDs), in whom respiratory insufficiency is the most common cause of premature mortality. Non-invasive or tracheostomy ventilator assistance has improved their quality of life and extended their survival [1, 2]. In contrast, patients with NMD are susceptible to the development of thoracic deformities such as scoliosis because of a combination of muscle weakness, spasticity and incompetent muscle control [3, 4]. This condition predisposes the trachea to be pinched between the innominate artery and the spine in the narrow superior mediastinum and to be compressed by the innominate artery.

Innominate artery compression of the trachea (IACT) is generally associated with persistent pulsatile contact between the anterior wall of the trachea and the adjacent innominate artery. This can form an erosion or granulation in the tracheal wall, leading to the development of tracheoinnominate artery fistula (TIF) and tracheomalacia. TIF is a fatal complication that usually presents with massive tracheal bleeding, often causing haemorrhagic shock and asphyxiation. Tracheomalacia is also a potentially life-threatening complication that leads to the development of collapsible tracheal obstruction, which often requires resuscitation. The major risk factor of these catastrophic IACT-related complications is a tracheostomy in which the cannula directly contacts the tracheal wall [5]. Because patients with NMD often require tracheostomy ventilation, they are considered to be at increased risk of developing of TIF and tracheomalacia following IACT [6].

To prevent TIF formation and exacerbation of tracheomalacia following IACT, we have recently reported a total of 10 cases of preventive innominate artery transection in patients with NMD...
Other investigators have also recently described preventive transection or ligation of the innominate artery [10-12]. Because no clear consensus on surgical treatment for IACT exists till date, the purposes of this study were to review our surgical management of IACT in patients with NMD and to assess the clinical benefits of preventive elective surgery.

PATIENTS AND METHODS

This study was performed with the approval of the institutional review board in both institutions, and the need for individual consent was waived. Clinical medical records and databases were retrospectively reviewed.

Patients

Between November 2002 and July 2012, 30 patients (19 males and 11 females) with NMD underwent surgical treatment for IACT in Kobe Children’s Hospital (23 patients) and Kobe University Hospital (7 patients). The patient demographics and clinical characteristics are summarized in Table 1. The median age at surgery was 12.8 years (range, 3.5-29.4 years) and the median body weight was 20.5 kg (range, 7.7-30.0 kg). All patients had inherited or acquired NMD, and 22 patients (73%) had thoracic deformities such as scoliosis. The progression of neurological or neuromuscular dysfunction required tracheostomy in 27 patients, laryngotracheal separation in 12, gastrostomy in 12 and fundoplication in 10 before surgery. Clinical characteristics and outcomes between elective surgery cases (Group A, n = 20) and emergent surgery cases (Group B, n = 10) were compared and analysed. The median postoperative follow-up period was 3.0 years (range, 0.4-10.2 years).

Preoperative assessment

All patients underwent preoperative tracheal assessment with bronchoscopy either routinely or after the onset of IACT-related clinical symptoms such as massive/minor tracheal bleeding, respiratory distress or obstructive apnoea. Pulsatile extrinsic compression of the anterior tracheal wall with/without erosion or granulation was noted in the latest bronchoscopic finding of each patient. Tracheomalacia around the anterior tracheal wall was observed in 16 patients. Twenty-eight patients underwent enhanced computed tomography (CT) of the chest before surgery. External compression of the anterior tracheal wall by the innominate artery was observed in 6 of the most recent patients since 2009, anatomical configuration around the trachea and the innominate artery were carefully outlined and evaluated by using three-dimensional multidetector-row CT angiography (3D-MDCTA). To assess cerebral circulation, such as a circle of Willis and other cerebral arteries, magnetic resonance angiography (MRA) or 3D-MDCTA of the brain was performed in 22 patients.

Operative procedure

The surgical strategy and techniques for treatment of IACT were identical in both institutions. The surgical management was performed by a co-operative team consisting of cardiovascular and paediatric surgeons. Appropriate skin incisions with or without sternotomy to expose the innominate artery were individually made (Fig. 1).

Innominate artery transection. The innominate artery was carefully isolated from the compressed tracheal site. After proximal and distal clamping of the innominate artery, the artery was ligated and transected. Both proximal and distal stumps of the innominate artery were closed with continuous 5-0 monofilament polypropylene sutures and were proximally and distally detached from the trachea after declamping the innominate artery. (Fig. 2A and B).

Innominate artery reimplantation. The innominate artery was clamped and transsected over the compressed tracheal site under full heparinization. The proximal end of the innominate artery was closed with continuous 5-0 monofilament polypropylene sutures.

![Figure 1: Surgical approach. (A) Skin incision: (a) I-shaped, (b) T-shaped, (c) inversed L-shaped, (d) curvilinear transverse. (B) Sternotomy: (a) full median sternotomy, (b) partial median sternotomy, (c) oblique partial manubriotomy.](https://academic.oup.com/ejcts/article-abstract/45/2/305/440727)
The right radial artery pressure (rRAP) was measured through curvilinear transverse skin incision without sternotomy. Elective surgery case before transaction. (C) Extra-anatomical innominate artery reimplantation using prosthetic vascular graft through I-shaped skin incision with full sternotomy. Elective surgery case before reimplantation (D) after reimplantation. BCA: brachiocephalic artery (innominate artery); RCCA: right common carotid artery; RSCA: right sub-clavian artery; Tr: trachea; TIF: tracheoinnominate artery fistula; Ao: aorta; inn.V: innominate vein; G: prosthetic vascular graft.

Figure 2: Intraoperative photographs. (A) Innominate artery transection through curvilinear transverse skin incision without sternotomy. Elective surgery case before transection. (B) Innominate artery transection through I-shaped skin incision with full sternotomy. Emergent surgery case after transection. (C) Extra-anatomical innominate artery reimplantation using prosthetic vascular graft through I-shaped skin incision with full sternotomy. Elective surgery case before reimplantation (D) after reimplantation. BCA: brachiocephalic artery (innominate artery); RCCA: right common carotid artery; RSCA: right sub-clavian artery; Tr: trachea; TIF: tracheoinnominate artery fistula; Ao: aorta; inn.V: innominate vein; G: prosthetic vascular graft.

The artery was then reimplemented to the proximal aorta on the right side, away from the trachea, by using an 8-mm prosthetic vascular graft with continuous 5-0 monofilament polypropylene sutures (Fig. 2C and D).

Tracheal wall repair. In patients with TIF formation, tracheal repair was performed following innominate artery transection or reimplantation. The site of the tracheal fistula was carefully exposed and granulation tissues around the TIF were clearly removed. The tracheal defect was directly closed with interrupted sutures using 4-0 polydioxanone.

Intraoperative assessment

To evaluate the integrity of cerebral circulation in the surgical treatment of IACT, test clamping of the innominate artery during surgery was performed since 2004. Fifteen patients were monitored for transcranial regional tissue oxygen saturation (rSO₂) using near-infrared spectroscopy, with an INVOS 5100C cerebral oximeter (Somanetics, Troy, MI, USA). Due to the variation in baseline rSO₂ values between patients, a baseline rSO₂ should be determined for each patient before induction of general anaesthesia. Cerebral ischaemia was detected on the basis of the percentage rSO₂ reduction or absolute rSO₂ value by measuring rSO₂ in the right hemisphere at test clamping of the innominate artery: % rSO₂ reduction = (baseline rSO₂ - test clamping rSO₂)/baseline rSO₂ × 100. The right radial artery pressure (rRAP) was measured in 18 patients before and after test clamping, and the percentage rRAP reduction was calculated as follows: % rRAP reduction = (rRAP before clamping - rRAP after clamping)/rRAP before clamping × 100. Flexible or rigid bronchoscopy to evaluate the tracheal lumen was performed after induction of general anaesthesia and just after surgery.

Table 2: Patient demographics and preoperative clinical symptoms and assessments in each group

<table>
<thead>
<tr>
<th>Variables</th>
<th>Group A (n = 20)</th>
<th>Group B (n = 10)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender (M/F)</td>
<td>14/6</td>
<td>5/5</td>
</tr>
<tr>
<td>Age at surgery (years)</td>
<td>12.9</td>
<td>12.4</td>
</tr>
<tr>
<td>Body weight at surgery (kg)</td>
<td>19.5 (10.0–28.8)</td>
<td>23.0 (7.7–30.0)</td>
</tr>
<tr>
<td>Tracheostomy (n)</td>
<td>17 (85%)</td>
<td>10 (100%)</td>
</tr>
<tr>
<td>Duration from tracheostomy until surgery (years)</td>
<td>2.6 (0.2–14.8)</td>
<td>1.3 (0.2–25.0)</td>
</tr>
<tr>
<td>Preoperative clinical symptoms</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Massive tracheal bleeding (n)</td>
<td>0</td>
<td>10</td>
</tr>
<tr>
<td>Minor tracheal bleeding (n)</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>Respiratory distress (n)</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>Obstructive apnoea (n)</td>
<td>3</td>
<td>0</td>
</tr>
<tr>
<td>Pre- and intraoperative assessments</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chest enhanced CT (n)</td>
<td>20 (100%)</td>
<td>8 (80%)</td>
</tr>
<tr>
<td>Bronchoscopy (n)</td>
<td>20 (100%)</td>
<td>10 (100%)</td>
</tr>
<tr>
<td>Brain MRA/enhanced 3D-MDCTA (n)</td>
<td>18 (90%)*</td>
<td>4 (40%)*</td>
</tr>
<tr>
<td>Transcranial rSO₂ (n)</td>
<td>14 (70%)*</td>
<td>2 (20%)*</td>
</tr>
<tr>
<td>rRAP (n)</td>
<td>16 (80%)*</td>
<td>2 (20%)*</td>
</tr>
</tbody>
</table>

*P < 0.05 vs Group B.

Statistical analysis

Continuous values were expressed as means ± standard deviation for normally distributed variables and as medians with range for non-normally distributed variables. Continuous and categorical variables in the two groups were compared with unpaired Student’s t-test, Mann–Whitney U-test or χ² test as appropriate. All data were analysed using the GraphPad Prism software 5.0 (GraphPad Software, Inc., San Diego, CA, USA). A value of P < 0.05 was considered to be statistically significant.

RESULTS

Patient demographics and preoperative clinical symptoms and assessments in each group are summarized in Table 2. Before surgery, IACT-related clinical symptoms were observed in 25 patients (83%). Fifteen patients in Group A had minor tracheal bleeding, respiratory distress or obstructive apnoea, whereas all patients in Group B had massive tracheal bleeding that required temporary over-inflation of a cuffed endotracheal tube at the TIF level. Almost all patients in both groups underwent enhanced CT of the chest and bronchoscopy on at least one occasion to assess the trachea before surgery. The number of patients who underwent preoperative MRA/enhanced 3D-MDCTA of the brain was significantly higher in Group A than in Group B (18 vs 4 patients: P = 0.004). Four of the 18 patients who underwent MRA assessment for cerebral circulation in Group A had abnormal findings such as incomplete or unclear circle of Willis: a hypoplastic circle of Willis in 2 patients, an incomplete circle of Willis with posterior...
transverse skin incisions were made significantly more often in Group A than in Group B (17 vs 2 patients; \( P = 0.019 \)). The baseline rSO2 value of the right cerebral hemisphere before test clamping was 83.9 ± 9.3. After clamping the innominate artery, the absolute rSO2 value was 81.5 ± 11.6, and the percentage rSO2 reduction by test clamping was 3.2 ± 4.9% from baseline (range, 0–14.5). The rRAP was also measured in a higher number of patients in Group A than in Group B (16 vs 2 patients; \( P = 0.002 \)). The rRAP decreased from 83.3 ± 13.7 to 55.4 ± 7.0 mmHg by test clamping. The percentage rRAP reduction was 32.4 ± 9.4% (range, 16.7–45.0). In all the four patients who had abnormal MRA findings of their circle of Willis, the percentage rSO2 and rRAP reduction were within threshold limit values. Three of these patients underwent innominate artery reimplantation based on their abnormal MRA findings, whereas the fourth patient underwent innominate artery transection based on the acceptable percentage rSO2 reduction.

### Operative and postoperative outcomes

The operative procedures in each group are summarized in Table 3. In Group A, 17 of 20 patients underwent innominate artery transection, and the remaining 3 patients underwent innominate artery reimplantation. In Group B, all the 10 patients underwent innominate artery transection. Skin incisions were included I-shaped in 5 patients, T-shaped in 12, inverted L-shaped in 3 and curvilinear transverse incision in 10. Curvilinear transverse skin incisions were made significantly more often in Group A than in Group B. The approach to the innominate artery was obtained through full median sternotomy in 7 patients, through partial median sternotomy in 4 (limited to the upper third in 10 or the upper half in 4) or through oblique partial manubriotomy in 1. Eight patients in Group A underwent innominate artery transection without sternotomy. For 1 patient in Group A, curvilinear transverse incision without sternotomy was converted to a T-shaped incision with partial median sternotomy because of unexpected intraoperative bleeding from the innominate artery. Tracheal wall repair for the TIF was performed in 7 patients in Group B. One patient in Group A underwent concomitant tracheoplasty for severe tracheomalacia. Bronchoscopy just after surgery produced complete relief of the pulsatile extrinsic compression in the anterior tracheal wall in all patients. Operative and postoperative outcomes in each group are shown in Fig. 3. The operative time was significantly shorter in Group A than in Group B (139 ± 52 vs 187 ± 59 min; \( P = 0.019 \)), and the operative blood loss in Group A was significantly less than that in Group B (69 ± 118 vs 303 ± 223 ml; \( P = 0.004 \)). No blood transfusion was required in Group A. Hospital stay after surgery was significantly shorter in Group A than in Group B (16.7 ± 14.7 vs 30.5 ± 14.4 days; \( P = 0.011 \)). No operative or early death occurred in any patients. Late death occurred in 3 patients in Group A: however, none of these patients was associated with the surgical treatment of IACT. No neurological complications such as brain infarction, bleeding or dysfunction occurred in any patients. The postoperative cognitive status remained the same as the preoperative status in all patients. None of the patients experienced symptoms of right upper extremity ischaemia or sub-clavian steal syndrome after surgery. Delayed wound healing was observed in 2 patients in Group A, but no wound infection or mediastinitis was observed after surgery. Three patients with severe preoperative tracheomalacia in Group A showed no improvement in malacic symptoms after surgery. Two of these patients underwent tracheostomy to insert a tracheostomy tube as an internal stent, and the remaining 1 patient underwent aortopexy and removal of the manubrium to relieve tracheal compression by the aortic arch. Of the 3 patients who underwent innominate artery reimplantation in Group A, 2 received postoperative antiplatelet therapy with aspirin and the remaining 1 patient did not receive any antiplatelet therapy. The prosthetic grafts in these 3 patients were all patent in the median postoperative follow-up period of 3.1 years (range, 3.0–8.2 years).

### DISCUSSION

The present study was a clinical review and assessment of our 10-year experience of surgical management of IACT in patients with NMD. The aim of our surgical management is to prevent...
life-threatening events, including massive bleeding or obstructive apnoea and improve the quality of the patient’s life.

The incidence of TIF has been estimated to be 0.1–1% after tracheostomy [13], but the true incidence of TIF in patients with NMD has not been reported till date. Although recent advances in respiratory support technologies has remarkably extended patient’s survival [1, 2], the development of thoracic deformity and the long-term placement of a tracheostomy tube can result in an increased incidence of TIF. The growing interests in the surgical management of IACT in Eastern Asia can be inferred by the recent high prevalence of publication from this area. The survival rate when massive bleeding of TIF occurs has been reported to be 14.3% [14]. This low survival rate seems to be attributable to the fact that it is difficult to control the unexpected bleeding to achieve haemodynamic and airway stability. The first manoeuvre should be the placement of a tracheostomy tube or an oral endotracheal tube with over-inflation of its cuff at the level of the bleeding point, and emergent surgery can then be performed as soon as the bleeding is under control. Early detection and diagnosis is the key to successful management of the massive bleeding of TIF. Nine of 10 patients undergoing emergent surgery in this study have already been suspected to have IACT by routine tracheal assessment on enhanced CT of the chest or bronchoscopy before the fatal event could occur. Their bleeding was immediately controlled by over-inflation of the cuff of the tracheostomy tube by caregivers, ambulance teams, nurses or doctors. As a result, none of them expired or had neurological insult despite massive bleeding. However, the remaining 1 patient who underwent emergent surgery went into cardiac arrest soon after admission and received cardiopulmonary resuscitation. Although there were no observed differences in time-related outcomes such as mortality and morbidity between the two groups in the present study, a life-threatening event should be avoided to prevent deterioration of the quality of life in patients with NMD.

Various therapeutic options for IACT have been previously reported [7–12, 15–18]. Although none of the therapeutic options for primary (intrinsic) tracheomalacia has gained widespread acceptance for its efficacy [19], the fundamental therapy for secondary (extrinsic) tracheomalacia due to IACT would be a complete release of the causal external compression of the innominate artery from the trachea. Aortopexy itself would not be effective for IACT because of the narrow superior mediastinum in patients with NMD [8]. The developed thoracic deformity and the long-term placement of a tracheostomy tube may exacerbate IACT again after surgery. Therefore, we strongly recommend innominate artery transection or reimplantation as preventive elective surgery in patients with NMD.

One of the biggest benefits of preventive elective surgery for IACT is the avoidance of fatal tracheal bleeding. Once massive bleeding due to TIF occurs, the lives of patients with NMD are suddenly put at risk and emergent innominate artery transection should be immediately performed without detailed assessments of the chest and brain. Insufficient brain assessment runs the potential risk of ischaemic brain damage secondary to emergent surgery. The present study demonstrated that preventive elective surgery for IACT results in significantly less operative blood loss and no blood transfusion compared with emergent surgery. Excessive bleeding affects outcomes adversely because it is associated with haemodynamic instability, prolonged operative time and increased need for allogeneic transfusion. Blood transfusion carries a risk of transmissions of infections and immunological reactions that cause organ dysfunction [20]. Therefore, surgery with less blood loss and no blood transfusion has great advantages for patients with NMD to improve postoperative outcomes by decreasing morbidity and mortality.

The other benefits of prophylactic elective surgery for IACT are that it is a minimally invasive procedure and involves a smaller skin incision, more limited sternotomy and shorter operative time. Sternotomy and prior tracheostomy generally impose a considerable risk for mediastinal infection in patients [21]. For patients with tracheostomy, minimally invasive surgery such as small skin incision and limited sternotomy has a great advantage in decreasing the risk of postoperative mediastinitis. In addition, the limited sternotomy can also be associated with sternal stability resulting in less postoperative pain, earlier recovery of respiratory function and shorter hospital stay. However, limited access and insufficient exposure of the operative field generate the risk of difficulty in controlling unexpected operative bleeding. Safe and reliable surgery is indispensable even if it is a minimally invasive procedure. One patient in this study experienced unexpected operative bleeding from the innominate artery, but conversion to a full sternotomy to control the bleeding was easily accomplished in an expeditious manner. Because there can be considerable variation in the anatomical relationships of the innominate artery and trachea in patients with NMD, it is quite important to plan the surgical approach based on a full understanding of the anatomical relationship. The preoperative 3D-MDCTA evaluation can provide surgeons with the information necessary to determine the optimal surgical approach and procedure.

The prevention of exacerbation of tracheomalacia is also one of benefits of preventive elective surgery for IACT. In the present study, 16 patients (53%) had tracheomalacia around the anterior tracheal wall, and clinical respiratory symptoms, including respiratory distress or obstructive apnoea, were observed in 9 patients (30%) before surgery. Most of the patients had improved respiratory symptoms or maintained their respiratory condition without an exacerbation of tracheomalacia after preventive elective surgery. However, 3 patients who had experienced obstructive apnoea before surgery continued to experience severe tracheomalacia and needed tracheostomy or aortopexy after surgery. Their timing of the preventive elective surgery should ideally have been earlier, before the progression to severe tracheomalacia.

The major concerns of innominate artery transection are cerebral circulation and neurological complications after surgery. Generally, adequate cerebral blood flow can be maintained by the circle of Willis and the numerous collateral pathways that redistribute blood to the deprived side in patients with unilateral internal carotid artery occlusion [22]. However, a recent report demonstrated that there may be some variations in the circle of Willis, such as absence, hypoplasia, duplicate or aneurysmal formation, in 54.8% of medicolegal autopsies [23]. We assessed cerebral circulation by MRA or 3D-MDCTA of the brain before surgery, and monitored transcranial rSO2 and rRAP during surgery. These assessments of cerebral circulation can be used as a basis on which selection of innominate artery transection or reimplantation is performed to maintain adequate cerebral circulation. In this series, the elective innominate artery reimplantation was performed in 3 patients who had incomplete or hypoplastic circle of Willis, as judged by preoperative abnormal image findings as the most important issue. Critical values of transcranial rSO2 were generally defined as a decrease of >20% from baseline or an rSO2 absolute value of <50%, which could be associated with an increase in neurological damage and/or cognitive impairment [24, 25]. Based on the evidence of transcranial rSO2, we conducted
innominate artery transection in 1 patient who had a stenotic segment of middle cerebral artery. The patient’s percentage rSO2 reduction and absolute rSO2 value was 4.8% and 63, respectively; both of these values were considered to be permissive for adequate cerebral circulation. The patient had no resultant neurological manifestations after surgery. With regard to the rRAP reduction in test clamping of the innominate artery, no clinical evidence of critical values for neurological damage has been reported till date. At present, we define a safe value for percentage rRAP reduction as a decrease <40% of rRAP before clamping. Further studies of percentage rRAP reduction would be necessary for an assessment of cerebral circulation.

Based on the results of the present study, our surgical and decision-making strategies for IACT in patients with NMD is shown in Fig. 4. Routine tracheal assessment on enhanced CT/3D-MDCTA of the chest and bronchoscopy is important for related clinical symptoms, surgical treatment is strongly advisable TIF or exacerbation of tracheomalacia. In patients with IACT-associated with IACT-related clinical symptoms and develop into gently undergo bronchoscopy, because these symptoms may be bleedings such as haemoptysis and blood on suction should ur- gerently undergo bronchoscopy, because these symptoms may be associated with IACT-related clinical symptoms and develop into TIF or exacerbation of tracheomalacia. In patients with IACT-related clinical symptoms, surgical treatment is strongly advisable when bronchoscopic findings demonstrate that the patient’s trachea underlying the innominate artery has persistent pulsatile compression on the anterior wall, with luminal malacic flattening and narrowing. To determine the optimal operative procedure, anatomical configuration of the trachea and innominate artery in the mediastinum should be evaluated using enhanced CT/3D-MDCTA of the chest. Innominate artery transection should be considered when the circle of Willis and other cerebral arteries are normal on brain MRA/3D-MDCTA findings. If these are not normal, innominate artery reimplantation should be considered. At present, we plan to perform innominate artery reimplantation only when a significant increase in percentage of rSO2 reduction (>20% from baseline) or a significant decrease in absolute rSO2 value (<50) in test clamping of the innominate artery is found in patients with NMD who have abnormal finding of cerebral circulation on brain MRA/3D-MDCTA findings.

In conclusion, preventive elective surgery for IACT (innominate artery transection or reimplantation) provides many advantages, including minimally invasive procedures and successful post-operative outcomes without neurological complications in patients with NMD. When the procedure is scheduled and performed in accordance with the optimal surgical strategy, including cerebral circulation assessment, this preventive elective surgery would be an optimal treatment for IACT not only to prevent fatal complications but also to improve the quality of life in patients with NMD.

Conflict of interest: none declared.

REFERENCES


Figure 4: Algorithm for the surgical management of IACT in patients with NMD.