

Airway Compression in Children With Congenital Heart Disease Evaluated Using Computed Tomography

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Background. Extrinsic airway compression often complicates the course of congenital heart disease (CHD) repair. This study investigated the risk factors and outcome of airway compression evaluated using computed tomography (CT) in CHD patients.

Methods. Of the 2,729 patients who underwent heart surgery for CHD between 1999 and 2007, airway compression was confirmed using CT in 58 (2.1%) patients. The patients were divided into groups according to the underlying CHD, and their medical records and CT scans were reviewed retrospectively.

Results. Airway compression was found more frequently in the vascular ring or absent pulmonary valve syndrome (8 of 11) and repaired aortic arch (22 of 213) groups than in the other groups (28 of 2,505) ($p < 0.001$). Patients with more severe respiratory manifestations showed greater airway compression on CT ($p < 0.001$) and

had a higher rate of additional surgery to relieve airway compression using multivariate analysis ($p = 0.005$). Airway compression was ameliorated in 13 of 17 patients after surgery for airway compression. Funnel chest deformity worsened after aortic arch repair and was associated with the need for surgical relief of airway compression. Pulmonary overflow disease could be followed up without additional surgery for airway compression.

Conclusions. Early airway compression detection and management may reduce further morbidity, especially after aortic arch repair. The patient's respiratory manifestation and the underlying disease characteristics must be considered when determining the need for additional surgery for airway compression.

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Respiratory problems ranging from abnormal findings on chest radiography to severe respiratory difficulty are common in patients with congenital heart disease (CHD). Extrinsic airway compression (AC) must be considered in the presence of respiratory insufficiency [1]. The AC commonly occurs in patients with anomalies of the aortic arch system and vascular rings. However, several underlying conditions cause AC in preoperative CHD cases, including a left-to-right shunt causing dilatation of the pulmonary arteries, truncus arteriosus [2], tetralogy of Fallot with an absent pulmonary valve, and a malpositioned or dilated aorta. The AC can even occur after surgeries for CHD such as the arterial switch operation, repair of an interrupted aortic arch or truncus arteriosus, and conduit reconstruction of the pulmonary outflow tract [3, 4]. Because chronic AC in childhood has significant morbidity and mortality, a definitive diagnosis and proper management are essential. To elucidate the clinical course of patients with AC and to investigate high-risk conditions

that require surgery for AC relief, we evaluated CHD patients with airway narrowing using computed tomography (CT) imaging and analyzed the CT findings.

Patients and Methods

Of the 2,729 patients who underwent heart surgery, open or closed, for CHD at Seoul National University Children's Hospital, Seoul, Korea between 1999 and 2007, external AC was demonstrated on cardiac or chest CT in 58 patients. The CT exams were performed in 483 patients (17.7%) with CHD for cardiovascular or airway imaging during the study period. Computed tomography was selected for AC evaluation because other imaging methods were rarely performed during the study period in our institution. Exclusion criteria were as follows: isolated congenital tracheal stenosis; airway narrowing due to prolonged endotracheal intubation; tracheoesophageal fistula; coronary artery disease; and cardiomyopathy. Patients with an intrinsic bronchus defect and primary or secondary endobronchial lesions were not included. Medical records and CT images were reviewed with regard to airway symptoms, underlying diseases, and surgical management.

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Abbreviations and Acronyms

AA group	= repaired aortic arch group
AC	= airway compression
AC ratio	= ratio of the compressed airway diameter to the trachea
Additional SAC	= SAC after first surgery for underlying heart disease
CHD	= congenital heart disease
CT	= computed tomography
FC ratio	= funnel chest deformity ratio
Group 1	= without SAC
Group 2	= with SAC (2A = simultaneous, 2B = additional)
IA ratio	= interaortic distance ratio
LRS group	= pulmonary overflow disease (left to right shunt) group
RVOT group	= reconstruction of the right ventricular outflow tract group
SAC	= surgery for airway compression
VR group	= vascular ring or absent pulmonary valve syndrome group

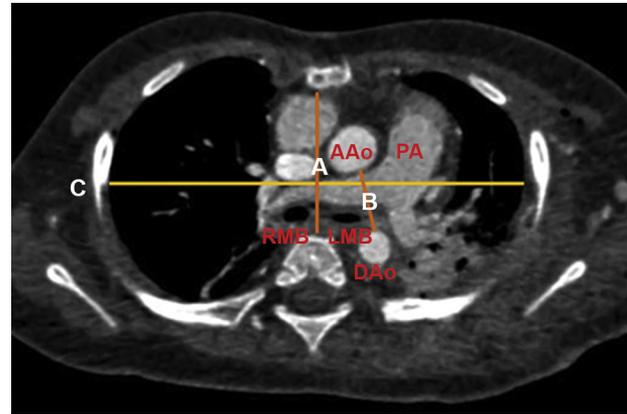


Fig 1. Airway evaluation on axial computed tomographic image. (A = length of sternum to spine; B = interaortic distance; C = longest lateral distance of the chest wall; A/C = funnel chest deformity ratio; B/C = interaortic distance ratio; AAo = ascending aorta; Dao = descending aorta; PA = pulmonary artery; LMB = left main bronchus; RMB = right main bronchus.)

Chest CT Analysis

Airway compression was defined as focal or diffuse narrowing from the trachea to the bronchi with evidence of external compression on CT. The ratios of the compressed airway diameter to the trachea (AC ratio), the interaortic (IA) distance to the length from the sternum to the spine (IA ratio), and the shortest anterior to posterior length (sternum to spine) to the longest lateral distance of the chest wall (funnel chest [FC] deformity ratio) were evaluated on axial CT images at the level of the proximal right pulmonary artery [5] (Fig 1). The AC severity was evaluated using the AC ratio.

Computed tomographic images were obtained using a 16-row multidetector CT scanner (Somatom Sensation 16; Siemens Medical Solutions, Forchheim, Germany). The scanning parameters used for enhanced thoracic CT imaging were as follows: detector collimation, 0.75 mm; gantry rotation time, 0.5 seconds; slice thickness, 1 mm; reconstruction interval, 0.5 mm; and a soft-tissue or smooth reconstruction algorithm or kernel. The protocol for the injection of contrast medium (Ultravist 370; Schering, Berlin, Germany) and image acquisition timing were described in detail elsewhere [6]. The CT imaging was performed under conscious sedation using chloral hydrate, and a bag valve mask was used to provide

positive pressure ventilation if the patient had a breathing problem without respiratory support.

Categorization According to Disease

Patients were categorized into 5 groups according to the underlying CHD (Table 1). Vascular ring (VR; n = 5, 3 double aortic arch and 2 pulmonary artery slings) or absent pulmonary valve syndrome (n = 3) were grouped together as the VR group because these diseases have native extrinsic AC by vessels; therefore, the approaches for treating AC associated with these diseases are similar in terms of AC. The repaired aortic arch (AA) group included coarctation of the aorta (n = 16), interruption of the aortic arch (n = 4), and diseases requiring a Norwood or Damus-Kaye-Stansel type of surgery (n = 2). The reconstruction of the right ventricular outflow tract (RVOT) group included the Rastelli operation or right ventricular-pulmonary artery conduit implantation with or without angioplasty of the pulmonary arteries. The pulmonary overflow disease (left-to-right shunt [LRS]) group consisted mainly of left-to-right shunt lesions without the coexisting conditions of the VR, AA, and RVOT groups. The group denoted “others” mainly consisted of functional single ventricles, transposition of the great arteries with an intact ventricular septum, isolated valvular disease, and Ebstein anomaly. Closed-heart surgeries were performed in 4 patients with vascular rings and 1 patient with coarctation of the aorta.

Table 1. Categorization of Congenital Heart Disease

Variable	No.	VR	AA	RVOT	LRS	Others	
Total patients	2,729	11	213	282	1944	279	
AC(+)	58	8 (72.7%)	22 (10.3%)	8 (2.8%)	20 (1%)	0	<i>p</i> < 0.001
Additional SAC	10	2	7	0	1	0	

AA = repaired aortic arch group; AC(+) = airway compression by computed tomography; additional SAC = surgical procedure for airway compression after first surgery for underlying heart disease; LRS = pulmonary overflow disease group; RVOT = reconstruction of right ventricular outflow tract group; VR = vascular ring or absent pulmonary valve syndrome group.

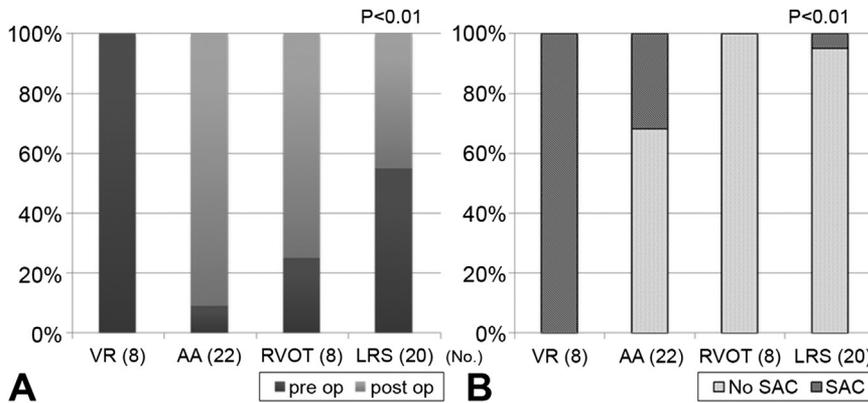


Fig 3. (A) Diagnosis of airway compression and (B) frequency of surgery for airway compression (concurrent or additional). (AA = repaired aortic arch group; LRS = pulmonary overflow disease group; No. = number of airway compression patients; Op = operation; RVOT, reconstruction of right ventricular outflow tract group; SAC, surgery for airway compression; VR = vascular ring or absent pulmonary valve syndrome group.)

Patients Treated With Additional SAC Versus Conservative Management

After the first surgery, with or without SAC, an additional SAC was performed for 10 (additional SAC [group 2B]) of the 58 patients for persistent or newly appearing AC in the 249 (range, 20 to 1699) days after the first operation. A severe respiratory manifestation grade was more frequent in the group 2B patients than in the conservative management group (without SAC [group 1]) (90% vs 8%, $p < 0.001$, Table 2). These group 2B patients required more respiratory support such as oxygen inhalation or mechanical ventilator support (66.6% vs 10.4%, $p < 0.001$) than the patients in group 1. The duration of endotracheal intubation and the intensive care unit stay were longer in group 2B patients than in group 1 patients (23.6 ± 21.2 days vs 8.9 ± 14.2 days [$p = 0.014$] and 24.8 ± 20.2 days vs 13.6 ± 16.1 days [$p = 0.07$], respectively). Pulmonary hypertension lasting longer than 1 month was more common in group 2B patients than in group 1 patients (40% vs 12.5%, $p = 0.036$). Funnel chest deformities were aggravated in all groups (FC ratio, 0.55 ± 0.09 to

0.49 ± 0.09 , $p < 0.001$), and a ratio of 0.5 or less was found in 90% of the group 2B patients and in 43.8% of the group 1 patients ($p = 0.008$). Using multivariate analysis, the respiratory support status (oxygen inhalation and mechanical ventilator support) was related to additional SAC ($p = 0.005$; odds ratio [OR] = 15.37; 95% confidence interval [CI], 2.23 to 105.72 [oxygen inhalation: OR = 2.7, CI = 0.4 to 17.6; mechanical ventilator support: OR = 31.3, CI = 2.9 to 328.6]).

Repaired Aortic Arch Group

Additional SAC was performed most frequently in the AA group (7 patients). In this group, AC was verified in 22 of a total of 213 patients (10.3%), and the FC ratio and IA ratio related to the airway space were changed. The FC deformities worsened after aortic arch repair (FC ratio, 0.59 ± 0.06 to 0.50 ± 0.06 ; $p = 0.001$), and this was more prominent in the patients who required additional SAC (Δ FC ratio, 0.11 vs 0.08, $p = 0.01$). The severity of the FC deformity had a statistically weak relationship with additional SAC ($p = 0.061$). Although the IA distance was shortened after aortic

Table 2. Comparison Between Conservative and Additional Surgery for Airway Compression Groups

Variable		Group 1 (Conservative, n = 48)	Group 2B (Additional SAC, n = 10)	p Value
Intubation duration (days)		8.9 ± 14.2	23.6 ± 21.2	0.014
ICU stay (days)		13.6 ± 16.1	24.8 ± 20.2	0.07
Respiratory manifestation grade	No	24	0	<0.001
	Mild	14	1	
	Moderate	6	0	
	Severe	4	9	
Respiratory support	Room air	43	3	<0.001
	On oxygen	4	2	
	On MV	1	4	
Pulmonary hypertension (>1 month)	No	42	6	0.036
	Yes	6	4	
Funnel chest deformity ratio	≤ 0.5	21	9	0.008
	> 0.5	27	1	

Group 1 = conservative management without surgery for airway compression (SAC); heart disease; ICU = intensive care unit; MV = mechanical ventilation.

Group 2 = additional SAC after first surgery for underlying

arch repair (IA ratio, 0.34 ± 0.07 to 0.30 ± 0.08 ; $p = 0.031$), this distance was not significantly different according to the additional SAC (Δ IA ratio, 0.08 vs 0.03; $p = 0.38$).

The additional SAC frequency differed according to the time of the aortic arch repair. The mean age of the patients at the time of the initial aortic arch repair was 33.5 ± 30.1 days (median, 20.5 [range, 6 to 129] days). In a comparison with early-repair (before 50 days of age) and late-repair (after 50 days of age) groups, additional SAC was required more frequently in late-repair patients than in early-repair patients (80% vs 20%; $p = 0.021$). However, changes in the FC and IA ratio after aortic arch repair were not significantly different according to the time of the arch repair. The procedures for the reconstruction of the right ventricular outflow tract (Rastelli operation or angioplasty of the pulmonary arteries) were not related to additional SAC in this AA group ($p = 0.29$).

Outcome

Freedom from additional SAC was significantly lower in the AA and VR groups than in the other groups ($p = 0.034$). Patients in the LRS group could be followed up without additional SAC (Fig 4).

During the follow-up period (4.2 ± 2.5 years) of 58 patients diagnosed with AC on CT, 3 patients died from non-airway problems, and 38% required hospitalization for respiratory distress with infection. In this study, 17% of patients underwent additional SAC and AC was relieved in all but 3 patients. Currently, 36 of 41 patients who did not undergo surgery for AC have had no symptoms associated with airway problems, but 2 patients in the AA group had persistent airway symptoms.

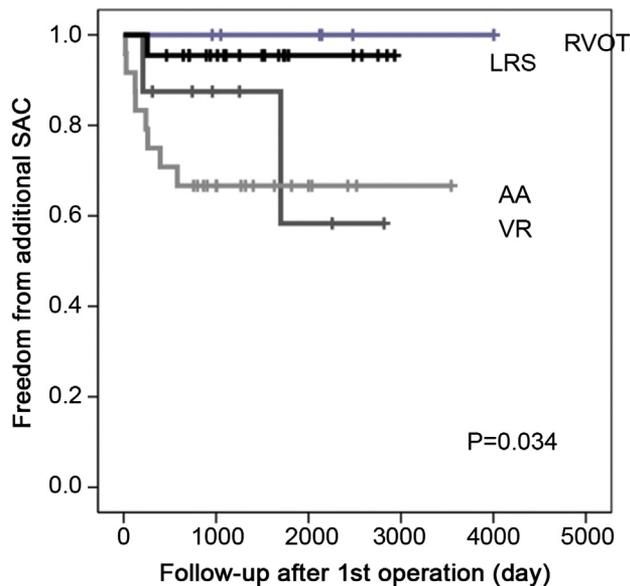


Fig 4. Freedom from additional surgery for airway compression. (AA = repaired aortic arch group; additional SAC = surgery for airway compression after first surgery for underlying heart disease; LRS = pulmonary overflow disease group; RVOT = reconstruction of right ventricular outflow tract group; VR = vascular ring or absent pulmonary valve syndrome group.)

Comment

There are many causes of respiratory problems in patients with CHD. The increased production of airway mucin in children who undergo cardiac surgery with cardiac pulmonary bypass is associated with respiratory complications [7]. The intrinsic instability of the airway wall, including tracheomalacia and bronchomalacia, affects the airway condition. Airway obstruction is among the factors attributable to extrinsic AC or congenital or acquired upper airway stenosis. Extrinsic AC is the most common cause of obstruction during the perioperative period of CHD [8]. In 2007, Efrati and colleagues [9] reported an extrinsic AC rate of 35.9% (14 of 39), which was detected by fiberoptic bronchoscopy before and after cardiac surgical repair in 39 patients with CHD and pulmonary abnormalities.

In infants, the airway is particularly vulnerable to extrinsic compression because the structures that support the airway are immature and airway conductance is low [10, 11]. In addition, these airways are prone to malacia even after surgical repair because of decreased resiliency [3]. Therefore, mild AC may lead to significant functional compromise. Airway compression frequently results in long-term ventilator dependency or recurrent respiratory problems [12]. Although the clinical presentations of AC include upper respiratory infection, wheezing, atelectasis, pneumonia, and hyperinflation, most symptoms are nonspecific and difficult to recognize. Therefore, more attention should be given to the timely recognition and early referral for the evaluation of AC to prevent long-term complications such as tracheobronchomalacia.

In this study it was difficult to predict AC based on the presence of respiratory symptoms because only 59% of patients with AC confirmed using CT had symptoms. On the other hand, greater AC was found with CT imaging when more severe symptoms were present; this suggests that in the presence of severe respiratory symptoms, aggressive AC evaluation is warranted.

Precise evaluation is important when determining the requirement of additional SAC, but magnetic resonance imaging has low accessibility and it is more expensive than CT examination in Korea. Although CT is an excellent noninvasive imaging modality that can be used to observe both the cardiovascular structure and the airway, CT is not permitted in all young CHD patients or minimally symptomatic patients because of the radiation hazard. In this study, the calculated effective dose from the dose length product values using a conversion coefficient of 0.021 for the pediatric chest was 0.62 (0.16 to 1.59) mSv [13]. These data are acceptable but it is better to avoid unnecessary radiation, especially in children. Therefore, the underlying disease, the grade of respiratory manifestations including the number of hospitalizations, and the status of respiratory support must be taken into consideration in place of the CT exam in order to avoid unnecessary surgery in cases in which spontaneous relief of AC is expected.

Aortic arch obstructions such as coarctation of the aorta or an interrupted aortic arch have a high probability of

causing abnormal geometry of the aortopulmonary space and AC [1, 14-16]. The procedure for repairing aortic arch obstructions may cause AC by relatively shortening the space between the ascending and descending aorta or by decreasing the aortopulmonary space. Jhang and colleagues [12] reported that AC was newly detected after aortic arch repair in 33.3% of patients in whom AC was not detected before surgery with or without a CT exam. In this study AC was common in the AA group, it was mostly detected after arch repair, and additional SAC was carried out more frequently in the AA group than in the other groups. In addition, unfavorable structural changes around the airway, such as aggravation of the FC deformity and shortening of the IA distance, were observed after arch repair. However, a definite association was not found between the IA ratio and the additional SAC group. Thus, in AA patients the evaluation and treatment of AC should be considered in the presence of respiratory symptoms, especially after a repair procedure. In our institution the aortic arch repair technique to prevent AC is as follows: extensive dissection of the aortic branch and the descending thoracic aorta area is performed to decrease the tension of the anastomosis site, and aortic anastomosis is performed without foreign material in young patients whose tissue is regarded to be elastic enough. While patch angioplasty could be considered in elderly patients this did not occur in the present study.

Among internal or external airway stenting, aortoplasty, aortopexy, and slide tracheoplasty, which are surgical techniques for AC, optimal treatment should be based on the patient's condition and operative risk. Airway stenting is used for central airway compression. In patients with obvious external compression by a vessel supporting systemic circulation, an internal stent seems inadequate to relieve severe obstruction [11, 17].

This study has several limitations. First, because this study included AC cases identified only with CT imaging, potential AC cases may have been missed in patients who did not undergo cardiac or chest CT evaluations. False-positive or false-negative cases of AC on CT imaging could have affected the number of patients included. Second, dynamic evaluation of AC using bronchoscopy with different levels of positive pressure in the airways was not performed because CT was selected as the airway evaluation tool. Third, sedation methods and the level of positive pressure ventilation during CT exams were not controlled although they can influence the diameter of the airway on CT. Fourth, because it was a retrospective study, there was a limitation in grading respiratory difficulty; therefore, relatively objective indicators were selected.

In conclusion, additional SAC was required more frequently in the VR and AA groups and was associated with respiratory support requirement. The AC associated with LRS could be managed conservatively, and these kinds of lesions should be addressed with repair of the cardiac lesion without airway intervention. Early detection and proper treatment of AC may reduce further

morbidity in children, especially after aortic arch repair. It is important to consider the underlying disease and the respiratory manifestation status when determining the requirement of additional SAC.

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