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ORIGINAL ARTICLE

Non-operative management of congenital tracheal stenosis: criteria by computed tomography

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Abstract

Objectives Whether to perform surgical or conservative treatment for congenital tracheal stenosis (CTS) is controversial. Thus, the computed tomography (CT) criteria for conservative treatment of CTS were investigated.

Methods From 2005 to 2017, 28 CTS cases were included. The operative cases and preoperative death cases constituted the required intervention group (group I), and the non-operative surviving cases constituted the observation group (group O). The diameter of the tracheal narrowest part (DTNP) on CT was evaluated as a criterion for non-operative follow-up.

Results Chest CT was performed 19 times in 19 group I cases and 18 times in 9 group O cases. The median age of the patients that underwent CT scan examinations was 3.4 months (range 0–25 months) in group I and 22 months (range 0–60 months) in group O. The cut-off values of the non-operative criteria were 40.8% (AUC: 0.82, $p < .01$) normal for age of the trachea's narrowest part, and 41.6% normal for body weight (AUC: 0.92, $p < .01$), respectively.

Conclusions DTNP is 40% and more of the normal diameter appears necessary for non-surgical management. The present study suggests that the criteria for conservative management of CTS are that the DTNP is not less than 40% of the normal tracheal diameter, with a few symptoms.

Keywords Congenital tracheal stenosis · Computed tomography · Non-operative management · Criteria · Operative management

Introduction

Congenital tracheal stenosis (CTS) is a disorder with a tracheal membrane defect and complete tracheal rings. Affected patients present with a wide spectrum of airway symptoms. CTS is sometimes a potentially life-threatening condition because of varying degrees of airway obstruction. It is disquieting for some clinicians that the airway obstruction could progressively deteriorate as the child grows, and the child's lung would outgrow the capacity of tracheal air conductance, resulting in respiratory failure or death [1].

Surgical intervention is considered to offer the best chance of survival for children with CTS who have severe symptoms [2]. The surgery includes resection of the stenotic segment and primary anastomosis, patch tracheoplasty with cartilage, pericardium, or anterior wall of the esophagus, and tracheal dilatation with or without stent insertion. Most procedures have produced largely unfavorable results and are associated with very high morbidity and mortality rates [3].

The most recent technique, slide tracheoplasty, has been hailed as a revolution in the management of CTS, achieving significantly better results. However, this procedure is not an easy undertaking; it involves cardiopulmonary bypass (CPB) or extracorporeal membrane oxygenation (ECMO), both of which have their inherent associated complications and morbidity.

With the advent of bronchoscopy and other imaging studies, a form of CTS with less severe symptoms is being increasingly recognized [4]. Patient survival after conservative management has been reported previously [5]. Although CTS cases have complete tracheal rings, symptoms may

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disappear without surgery. Whether to perform surgical or conservative treatment is controversial. Conservative management may be more widely practiced than reported, yet the rationale behind it requires evidence.

The successful outcomes of a subgroup of conservatively managed patients with CTS are reported. In this study, the computed tomography (CT) criteria for selecting non-operative management of CTS cases were investigated.

Material and methods

Patient selection

A retrospective study of 28 consecutive cases of CTS managed in Shizuoka children's hospital between January 2005 and July 2017 was carried out. Subglottic stenosis, tracheomalacia, and bronchial stenosis were excluded from the series. The diagnosis of CTS was based on bronchoscopic and radiologic evidence, in addition to clinical findings. Overall, there were 28 cases of CTS: 17 operative cases, two preoperative death cases, and nine non-operative survival cases. The surgical cases and the preoperative death cases were defined as the required intervention group (group I), and the non-operative survived cases were defined as the observation group (group O), and they were compared. The control group consisted of 100 patients under 8 years of age without tracheal disease. The study was performed after approval from the institutional ethics committee (approval number of 2017105).

Data collection

Patients who have symptom of respiratory failure with resistance to medical treatment, or where the airway could not be secured for heart surgery were included in group I. The following patient data were collected: sex, gestational age, birth weight, age at presentation, weight at presentation, type of stenosis according to length, minimum diameter of the stenotic trachea, mortality, presence of tracheotomy, initial symptoms, current symptoms (or at time of tracheoplasty), follow-up, and comorbidities. The diameter of the tracheal narrowest part (DTNP) on CT was evaluated as a criterion for conservative management.

Multidetector-row CT (MDCT) examination

Patients were imaged in the supine position using a 64-slice CT scanner (Aquilion 16; Toshiba Corporation, Medical System Company**,** Tokyo, Japan). In patients older than 5 years, the scanning parameters were: collimation 0.75 mm, pitch 0.425, effective thickness 1.0 mm, reconstruction interval 1.0 mm, voltage 120 kVp, tube current 150–300 mAs, rotation time 0.50 s, and scan time 8–16 s. In patients younger than 5 years, they were: collimation 0.75 mm, pitch 0.50, effective slice thickness 1.0 mm, reconstruction interval 1.0 mm, voltage 120 kVp, tube current 100–150 mAs, rotation time 0.50 s, and scan time 4–8 s. Patients received 2.0 ml/kg of intravenous contrast medium (iopromidol 300; Nippon Schering, Osaka, Japan) for MDCT angiography using the antecubital vein at a rate of 4 ml/s through a 22-gauge catheter.

Evaluation of MDCT images and measurement of tracheal diameters

Two experienced reviewers assessed the MDCT images by consensus. All CT scans were evaluated for the presence of tracheal stenosis. All images were processed and measured using a soft-tissue window setting (width 400–450 HU; level 40–50 HU). The trachea was measured at the narrowest level of the trachea. The internal diameters of the trachea were measured.

In addition to the original 1.0-mm-thick axial slices, thick-slab maximum intensity projection (MIP) and other post-processing techniques such as multiplanar reconstruction (MPR) and three-dimensional (3-D) volume rendering were used, depending on the individual. These images were used to ensure that the tracheal axes were orthogonal to the central tracheal axis, thus avoiding oblique cut distortion (Fig. 1).

Operation (slide tracheoplasty)

After the induction of general anesthesia, standard median sternotomy was performed. The trachea was divided transversely through the middle of the stenosis, and the procedure then proceeded in the manner described by Grillo et al. [6]. After the procedure was completed, an anesthesiologist re-inserted the endotracheal tube under direct visual guidance. Patients were placed on the ventilator for at least 2 weeks to maintain airway stenting.

Non-operative management

Non-operative management consisted of treating respiratory tract infections, chest physiotherapy, and short-term oxygen therapy. Intervals between bronchoscopic and/or CT evaluations varied from 6 months for younger patients to 2 years for older asymptomatic patients. Patients were followed up until they stopped growing.

Theory/calculation

Categorical data are described using frequencies and percentages, and they were analyzed using the Chi-square and Fisher's exact tests. Continuous variables are described as

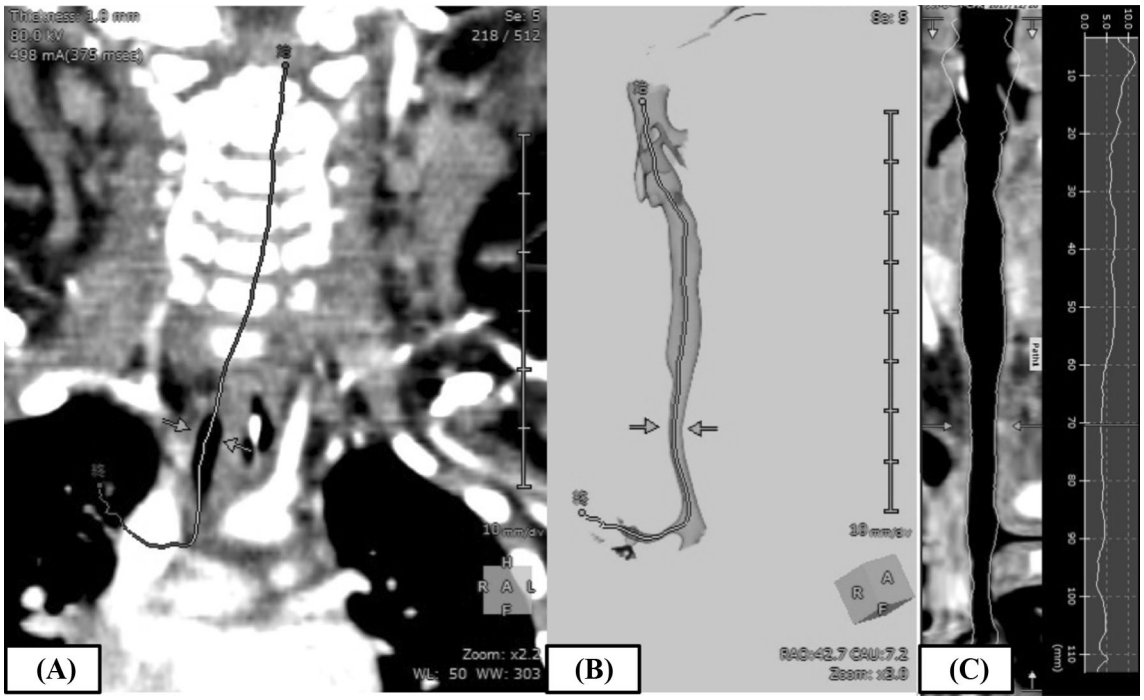

Fig. 1 Evaluation of MDCT images and measurement of tracheal diameters. Multiplanar reconstruction (MPR) and three-dimensional (3-D) volume rendering were used to ensure that the tracheal axes were orthogonal to the central tracheal axis, thus avoiding oblique cut distortion. **a** The coronal section; **b** the three-dimensional volume rendering; **c** the coronal section avoiding oblique cut distortion

medians and interquartile range (IQR) and compared using the Mann–Whitney *U* test. A receiver-operating characteristic (ROC) curve analysis was used to determine a suitable cut-off value for the DTNP. The accuracy or power of each variable for predicting survival was assessed by the area under the ROC curve (AUC), sensitivity, and specificity. All statistical analyses were performed using the JMP software program (version 12.01; SAS Institute, Inc, Cary, NC, USA). Values of $p < 0.05$ were considered significant.

Regression analysis for DTNP and age and weight between the control group, group I, and group O

The DTNP was investigated using 100 CTs of 100 children in the Control group, 19 CTs of 19 cases in group I, and 18 CTs of nine cases in group O. The relationships between the DTNP (mm; *y*-axis) and age and weight (in months and kilograms; *x*-axis) for the control group, group I, and group O were then investigated by regression analysis. Each regression line used least-squares approximation of the line to the points.

The observed/expected DTNP (o/e-DTNP)

The o/e-DTNP was expressed as a percentage of the appropriate mean DTNP for the 100 patients in the Control group. The o/e-DTNP was adjusted for body weight and age.

Results

Patient demographics

Congenital tracheal stenosis affected predominantly male infants (M/F = 16:13) in this series, and a congenital cardiac anomaly was the most common associated anomaly (66%, 19/29). Of 29 patients, 7 (24%) had an anomalous left pulmonary artery sling (Tables 1 and 2). The long-segment stenosis, where more than 50% of the tracheal length was stenotic, was the most common (76%). The patients presented initially with various symptoms of respiratory insufficiency just after birth ($n = 7$), respiratory insufficiency triggered by infection ($n = 5$), cyanosis at crying ($n = 5$), stridor ($n = 5$), during intubation for surgery ($n = 2$) or during intubation for examination ($n = 3$), and respiratory insufficiency with pneumothorax ($n = 1$) (Table 3). Median follow-up was 5 years. The group I patients presented relatively earlier than group O patients (1.5 vs 4.4 months), and their symptoms appeared more severe. The mortality rate of the group I (39%) was three times that of group O (11%) (Table 1). The median age of the patients that underwent CT scan examinations was 3.4 months (range 0–25 months) in group I and 22 months (range 0–60 months) in group O.

All operations were performed via median sternotomy under CPB except in four patients, who were approached through a low collar incision without CPB. All patients

Table 1 Patients' demographic data and outcomes

	Group I ($n=19$)	Group O ($n=9$)	p value
Sex (male: female)	8: 11	7: 2	.11
Gestational age, weeks, median (IQR)	37.4 (35–39)	37 (36–39)	.83
Birth weight, g, median (IQR)	2453 (1990–3204)	2352 (1813–2940)	.76
Age at presentation, months, median (IQR)	1.5 (0–5)	4.4 (0.3–15)	.36
Weight at presentation, kg, median (IQR)	3.3 (2.5–5.1)	6 (2.8–10)	.22
Type of stenosis by length			.62
Long-segment type	14 (74%)	8 (88%)	
Short-segment type	5 (26%)	1 (11%)	
Length of stenotic trachea, mm, median (IQR)	20 (12–28)	25 (21.5–35.5)	.06
DTNP, mm, median (IQR)	2.3 (1.6–2.5)	4.2 (2.9–4.6)	< .01*
o/e-DTNP by age, %, median (IQR)	37.6 (28.6–42.3)	47.8 (40.3–56.8)	< .01*
o/e-DTNP by weight, %, median (IQR)	37.5 (27.3–41.1)	49.3 (42.1–55.6)	< .01*
Mortality			.21
Preoperative death	2 (2 respiratory insufficiency)	–	
Postoperative death	5 (4 respiratory insufficiency, 1 heart failure)	–	
Sudden death	0	1 (unknown death)	
Tracheostomy	3	–	NA
Follow-up, years, median (IQR)	5 (1.5–8)	4 (2–6)	.75

DTNP diameter of the tracheal narrowest part, *o/e-DTNP* observed/expected diameter of the tracheal narrowest part, *IQR* interquartile range
*$p < 0.05$

Table 2 Associated congenital anomalies

	Group I ($n=19$)	Group O ($n=9$)
Cardiovascular anomalies	15 (79%)	4 (44%)
Left pulmonary artery sling	6 (32%)	1 (11%)
Patent ductus arteriosus	3 (16%)	0 (0%)
Atrial septal defect	2 (11%)	1 (11%)
Ventricular septal defect	6 (32%)	1 (11%)
Double outlet right ventricle	2 (11%)	1 (11%)
Pulmonary atresia with intact ventricular septum	1 (5%)	0 (0%)
Ebstein's malformation	1 (5%)	0 (0%)
Shone complex	1 (5%)	0 (0%)
Major aortopulmonary collateral artery	1 (5%)	0 (0%)
Right aortic arch	1 (5%)	0 (0%)
Unilateral pulmonary artery defect	1 (5%)	2 (22%)
Respiratory anomalies		
Single lung	1 (5%)	2 (22%)
Tracheal bronchus	3 (16%)	1 (11%)
Gastrointestinal anomalies		
Esophageal atresia	0 (0%)	1 (11%)
Anorectal malformation	2 (11%)	1 (11%)
Vertebral anomalies	1 (5%)	0 (0%)

underwent repair by slide tracheoplasty. In the 15 patients who had additional cardiac and vascular anomalies, six patients with cardiac anomalies and six patients with vascular anomalies were treated concomitantly. Two cases in group I died due to respiratory insufficiency while preparing for surgery. Group I contained more patients with symptoms that were more severe. The only case of mortality in group O was due to sudden unknown death;

Table 3 Initial symptoms

	Group I ($n=19$)	Group O ($n=9$)
Respiratory insufficiency just after birth	7 (37%)	0
Respiratory insufficiency triggered by infection	5 (26%)	0
Cyanosis at crying	4 (21%)	1 (11%)
Stridor	2 (11%)	3 (33%)
During intubation for surgery	1 (5%)	1 (11%)
During intubation for examination	0	3 (33%)
Respiratory insufficiency with pneumothorax	0	1 (11%)

the patient had CTS with complicated congenital heart disease.

Regression analysis for DTNP and age and weight between the control group, group I, and group O

In group I and group O, chest CT was performed 19 times in 19 cases and 18 times in nine cases, respectively. The relationships between the DTNP and age in months and weight (kg) are shown in Fig. 2 for the control group, group I, and group O.

Using age, the best fit for the DTNP in controls was represented by the regression line ($6+0.08\times$[age in months]) ($r=0.86$, $p<0.01$). The regression analysis also yielded the DTNP formulas ($2+0.03\times$[age in months]) ($r=0.36$, $p<0.01$) for group I and ($3+0.04\times$[age in months]) ($r=0.85$, $p<0.01$) for group O. Using weight, the best fit for the DTNP in controls was represented by the regression line ($4.7+0.35\times$[weight, kg]) ($r=0.91$, $p<0.01$). The regression analysis also yielded the DTNP formulas ($1.8+0.08\times$[weight, kg]) ($r=0.33$, $p<0.01$) for group I and ($2.7+0.12\times$[weight, kg]) ($r=0.75$, $p<0.01$) for group O.

Non-operative CT criteria

The o/e-DTNP by age was then expressed as a percentage of the expected mean for age and weight (o/e-DTNP by age $=4.7+0.35\times$[weight, kg]; o/e-DTNP by weight $=6+0.08\times$[age in months]).

An ROC curve analysis was used to compare the outcomes between those with DTNP measurements and the o/e-DTNP by age and weight outcomes.

The data in group I and group O are shown in Table 1. There were no significant differences between group I and group O in sex ($p=0.11$), gestational age ($p=0.83$), birth weight ($p=0.76$), age at presentation ($p=0.36$), weight at presentation ($p=0.22$), type of stenosis according to length ($p=0.62$), and length of the stenotic trachea ($p=0.06$) (Fig. 3).

According to the ROC curve analysis, the recommended cut-off values for the DTNP, the o/e-DTNP by age, and the o/e-DTNP by weight were 2.7 mm (sensitivity, 100%; specificity, 63%; AUC, 0.8; $p<0.01$), 40.8% (sensitivity, 63%; specificity, 84%; AUC, 0.82; $p<0.01$), and 41.6% (sensitivity, 89%; specificity, 84%; AUC, 0.92; $p<0.01$), respectively (Fig. 4).

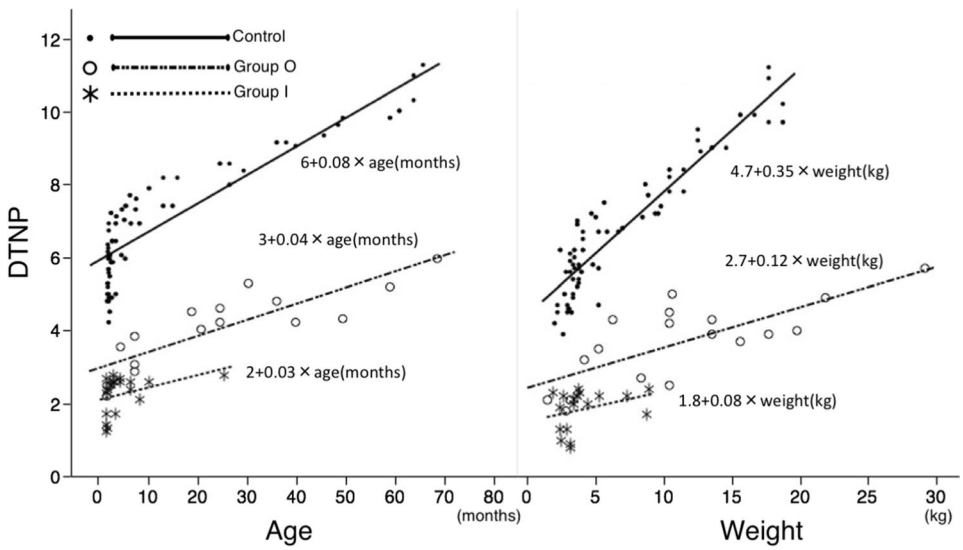

Fig. 2 Relationships between DTNP and age and weight in the control group, group O, and group I. The continuous line, dashed double-dotted line, and dotted line correspond, respectively, to the expected results for the control group, group O, and group I at each age and weight when diagnosed on CT. The dots, circles, and asterisks indicate the control group, group O, and group I, respectively. *DTNP* diameter of the tracheal narrowest part

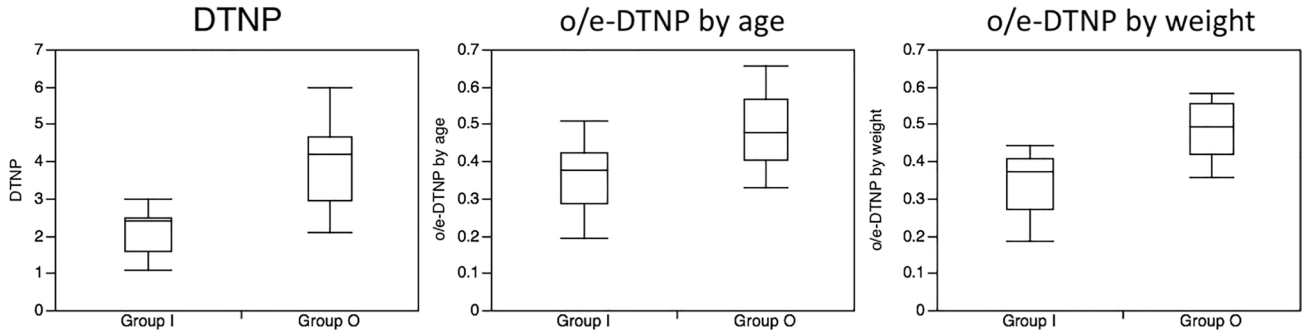

Fig. 3 Comparisons of DTNP, o/e-DTNP by age, and o/e-DTNP by weight between group I and group O. Box plots of the DTNP, o/e-DTNP by age, and o/e-DTNP by weight from group I and group O. The bars in the box correspond to median values, and the upper and lower bars correspond to the first and third quartiles, respectively. The two vertical lines outside the boxes indicate the maximal and minimal values. *DTNP* diameter of the tracheal narrowest part, *o/e-DTNP* observed/expected diameter of the tracheal narrowest part

Discussion

Various surgical procedures for CTS have been reported, and slide tracheoplasty for long-segment CTS, described by Tsang [7] and Grillo [6], has become a standard surgical procedure that has been shown to greatly improve outcomes. However, the mortality rate following tracheal surgery still reaches 10–85%, with an incidence of further complications rising up to 40%. Anastomotic leakage is one of the most dramatic complications, responsible for further tracheal re-interventions in from 28 to 48% of cases [8–10]. Chiu et al. showed that CTS repair is associated with the highest mortality in patients younger than 1 month and in patients with intracardiac anomalies [11]. This procedure is not without risk, and intervention is better justified in a patient with significant and worsening symptoms. Morita et al. reported that non-operative management should be attempted in patients with stable ventilation in the neonatal period [12].

The surgical correction of the mild forms of tracheal stenosis is still under debate. The concerns arise from the evidence that tracheal reconstruction by itself increases the risk of death or complications. Moreover, there is a growing number of reports showing encouraging results with conservative management [10, 13].

However, the standard for conservative management is also controversial. The standard in the present case was DTNP. In a previous study, computational flow dynamic studies of a web-like stenosis demonstrated that the degree of narrowing is the main determinant of the pressure drop across an airway's stenosis, rather than its length, and that the pressure drop correlates well with the symptoms [14].

In previous reports, DTNP of 3 mm was proposed by Huang et al. as a "cut-off" diameter above which left pulmonary artery repositioning could be performed alone [13].

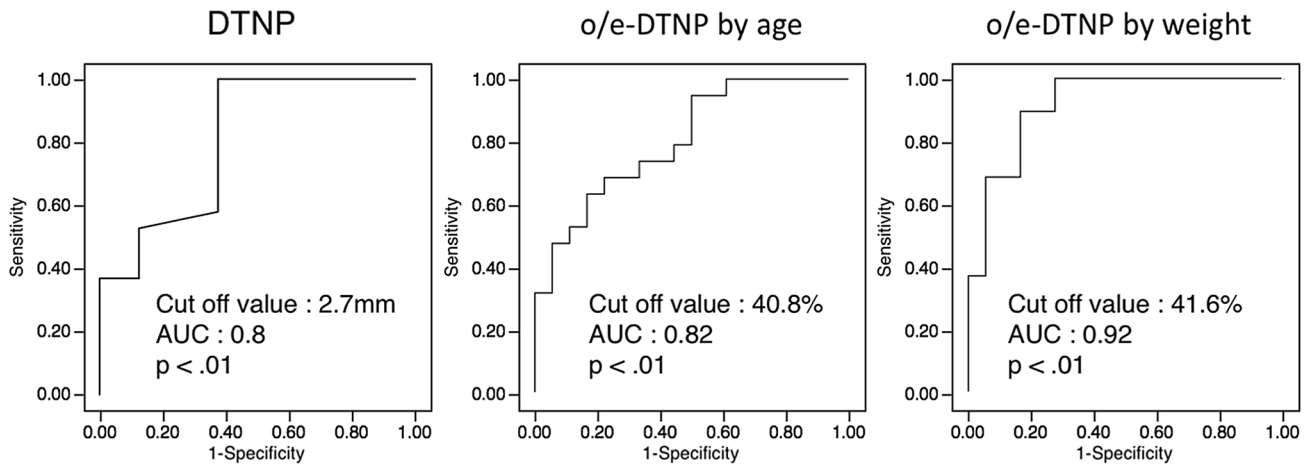

Fig. 4 ROC curve analysis for non-operative management with congenital tracheal stenosis by the DTNP values and by the o/e-DTNP (by age and weight) cut-off values. *AUC* area under the curve, *ROC* receiver-operating characteristic, *DTNP* diameter of tracheal narrowest part, *o/e-DTNP* observed/expected diameter of the tracheal narrowest part

Anton-Pacheco previously defined mild CTS for which conservative management was appropriate as DTNP of 4–6 mm with minimal clinical symptoms [15]. Morita et al. suggested that surgical intervention may be needed for a cases with a minimum diameter of the stenotic trachea of 2.0 mm or less in the neonatal period on CT [12].

Based on the results of the present study, the selection criteria for conservative management of CTS are as follows: (1) the DTNP is not less than 2.7 mm; and (2) the DTNP is not less than 40% of normal tracheal diameter, adjusted for weight and age. When the diameter is halved, the lumen area is reduced to one-fourth of that with a normal diameter, and the pressure dropped beyond the critical point in a flow dynamic study [14].

However, the question of whether the narrowed trachea (with complete rings) can grow to a normal diameter is still under debate [10, 13, 15, 16]. Cheng et al. showed growth of the trachea up to normal values for age [16]. Based on previously published observations [16, 17], they reported that a stenotic trachea, unlike scar tissue, is capable of growth. More importantly, the tracheal growth, and hence the tracheal lumen diameter enlargement, is faster than that of normal tracheal lumen growth, especially after infancy.

The long-term follow-up demonstrated that most of the conservatively managed children with CTS were symptom-free, with only two patients experiencing stridor. The present study demonstrated that conservative management is feasible and safe for these selected patients. As the tracheal airflow increase is proportional to the fourth power of the tracheal radius increase, the children's symptoms are expected to improve precipitously with tracheal growth.

This study has some limitations. First, it involved a small number of cases from a single tertiary pediatric center. Second, in group I and group O, chest CT was performed 19 times in 19 cases and 18 times in nine cases, respectively. In groups I and O, chest CT was performed once and one-to-three times, respectively. Finally, the DTNPs were measured using a soft-tissue window setting. If the DTNP were measured using a pulmonary window setting, the DTNP would appear smaller than the true value. Therefore, the present setting was width 400–450 HU, level 40–50 HU, but the value of DTNP changes according to the setting value.

Conclusions

The results of the present study suggest that the criteria for conservative management of CTS are that the DTNP is not less than 2.7 mm, and/or not less than 40% of the normal tracheal diameter, with a few symptoms.

However, infectious or inflammatory respiratory disorders, which reduce the tracheal lumen further, could cause apparent life-threatening events. In addition, later on, some patients may develop respiratory difficulties with exercise. In patients with persistent or worsening symptoms and a minimal increase in tracheal diameter, tracheoplasty is eventually required. A prospective comparison of various modalities of management would require prolonged follow-up. Careful and thorough work-up is warranted, and the treatment options of these patients have to be individualized.

Compliance with ethical standards

Conflict of interest None declare.

Ethical approval Clinical registration number: approval from the institutional ethics committee (approval number of 2017105).

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