

RESEARCH

Open Access



# Congenital heart diseases with airway stenosis: a predictive nomogram to risk-stratify patients without airway intervention

Qiyu He<sup>1†</sup>, Yuze Liu<sup>1†</sup>, Zheng Dou<sup>1</sup>, Kai Ma<sup>1</sup> and Shoujun Li<sup>1\*</sup>

## Abstract

**Background** This study focused on congenital heart disease (CHD) patients complicated with airway stenosis (AS) without airway intervention and aimed to identify the patients with potential risks.

**Methods** Patients diagnosed with CHD and AS were enrolled in this retrospective study. The primary outcome was defined as a postoperative mechanical ventilation duration of more than two weeks. We constructed a prediction model to predict the risk of prolonged mechanical ventilation (PMV).

**Results** A total of 185 patients diagnosed with CHD and AS in Fuwai Hospital from July 2009 to December 2022 were included in the study. Weight at CHD surgery, cardiopulmonary bypass (CPB) duration, complex CHD and comorbid tracheobronchomalacia were identified as risk factors and included in the model. The ROC curve showed a good distinguishing ability, with an AUC of 0.847 (95% CI: 0.786–0.908). According to the optimal cut-off value of the ROC curve, patients were divided into high- and low-risk groups, and the subsequent analysis showed significant differences in peri-operative characteristics and in-hospital deaths.

**Conclusions** With the predictive model, several factors could be used to assess the risky patients with PMV. More attention should be paid to these patients by early identification and routine surveillance.

**Keywords** Congenital heart diseases, Airway stenosis, Mechanical ventilation, Nomogram, Predictive model

## Background

Congenital heart disease (CHD) complicated with airway anomaly, a rare combination of lesions, approximately accounts for 3–4% of patients [1]. Tracheobronchial malacia, tracheal stenosis, and tracheoesophageal fistulas are the major presentations of the anomalies, and

surgical intervention may be needed occasionally [2, 3]. Airway stenosis (AS) is the major subtype and 50–70% of the patients are reported to have a concomitant cardiac anomaly [4–6].

Depending on the various presenting symptoms, different treatment strategies may be adopted. For severe conditions, surgical intervention may be needed. However, for AS patients with moderate respiratory symptoms, conservative management may be preferred. Cheng et al. indicated that compared to normal children, tracheal growth and tracheal diameter enlargement seem to be faster in congenital tracheal stenosis (CTS) children, especially after infancy [7]. In a large retrospective analysis of the Society of Thoracic Surgeons Congenital Heart

<sup>†</sup>Qiyu He and Yuze Liu contributed equally to this work.

\*Correspondence:

Shoujun Li

drlishoujunfw@163.com

<sup>1</sup>Pediatric Cardiac Surgery Centre, National Centre for Cardiovascular Diseases, State Key Laboratory of Cardiovascular Disease, Fuwai Hospital, Chinese Academy of Medical Sciences, Peking Union Medical College, Beijing 100037, China



Surgery Database (STS-CHSD), 6861 (3.4%) patients were complicated with airway anomalies and only 428 (0.2%) patients underwent tracheal operations during the same hospitalization [8].

Most of the studies focused on CHD patients who underwent tracheal surgery, so the data on the conservative group are scarce [9, 10]. As we mentioned, most CHD patients with tracheal anomalies may not require tracheal intervention, thus, studies exploring these populations are warranted to better understand the outcomes and prognosis. Consequently, in this study, we aim to retrospectively investigate the CHD patients who underwent conservative management of AS in our center, to provide novel insight to AS management.

## Methods

### Patient population and management

Patients with the diagnoses of CHD complicated with AS in Fuwai Hospital were reviewed from July 2009 to December 2022. For patients diagnosed with AS and CHD, computerized tomography (CT) scan or fiberoptic bronchoscopy would be prescribed. After adequate multidisciplinary team (MDT) consultation and preoperative evaluation by cardiac surgeons, cardiologists, echocardiologists, and radiologists, patients with stenotic airway segment detected by fiberoptic bronchoscopy or CT scan and significant respiratory symptoms would undergo tracheal surgery, such as slide tracheoplasty. However, in patients with AS reported by bronchoscopy or CT but mild or asymptomatic respiratory symptoms, conservative treatment on AS would be preferred. Consequently, we enrolled patients who met the following criteria: diagnosis of CHD, AS determined by fiberoptic bronchoscopy or CT scan, no significant respiratory symptom, and no tracheal surgery during hospitalization.

### Definitions

The Society of Thoracic Surgeons- European Association for Cardiothoracic Surgery (STAT) mortality category was selected to risk-stratify the CHD procedures. Complex CHD was defined as CHDs excluding simple shunts (atrial septal defect, ventricular septal defect and patent ductus arteriosus), pulmonary artery slings and vascular rings [11]. The primary outcome was defined as prolonged mechanical ventilation (PMV). According to previous reports and our clinical experience, PMV was defined as postoperative mechanical ventilation for more than 14 days [12–14].

### Statistical analysis

Descriptive analysis of baseline data was presented by median (interquartile range, IQR) for continuous variables, or frequency (percentage) for categorical variables. Wilcoxon rank sum test were used to compare the

continuous data and the chi-square test or Fisher exact test was used to analyze categorical data. Variables with missing values were detected and processed by the Mice package of R. The imputation methods were selected based on the type of variables that continuous data were imputed using Predictive Mean Matching, and categorical data were imputed using Logistic Regression Imputation. The imputed dataset was compared with the original dataset to verify the bias generated in the process. The imputed dataset was used for the following statistical analysis.

Logistic regression was used to assess the association between perioperative indicators and prognosis. Variables with potential predictive power were first included in the univariate logistic regression analysis, and those with  $P < 0.1$  were further included in the multivariate logistic regression analysis.

We tested the accuracy of the model with the Hosmer-Lemeshow test and presented it with calibration plots. The receiver operating characteristic (ROC) curve was plotted to determine the model's ability to accurately identify PMV in children with CHD undergoing conservative treatment of AS. We also identified the net benefit of the model by decision curve analysis (DCA). Finally, a nomogram was adopted to visualize the model.

Due to the lack of an external validation cohort, we internally validated the model using machine learning method. Cross-validation was selected to examine the accuracy and analyze the overfitting of the predictive model. After randomly divided all the data in the original cohort into  $k$  groups,  $(k-1)$  of them were taken as the training cohort for constructing the model, and the remaining 1 group was treated as the validation cohort ( $k$ -fold cross-validation). The model constructed from the training cohort was used to predict the outcome of the validation cohort. To reduce the influence of random grouping, the above process was repeated 200 times. The other method we used for internal validation was bootstrap validation. The training cohort was constructed by sampling (with replacement) from the original cohort. Although the probability of each patient being selected was the same, the training cohort differed significantly from the original cohort due to the use of sampling with replacement. Similarly, the process was repeated 200 times. We used these two methods to obtain the corrected area under curve (AUC) values and 95% confidence intervals for model correction, respectively.

Based on the results of logistic regression, the possible risk score for each patient was calculated, and the patients were divided into high-risk and low-risk groups according to the optimal cutoff value of the ROC curve. We further compared the differences in other postoperative indicators between the two groups, including

postoperative hospital stay, ICU stay, reintubation, ECMO implication, and in-hospital death.

All the statistical analyses were performed using R-studio (version 4.2.2),  $P < 0.05$  was considered to be statistical significance.

## Results

### Baseline characteristics

This retrospective study comprised 221 patients diagnosed with CHD and AS, of which 36 patients underwent airway surgery (15 with slide tracheoplasty, 21 with tracheobronchial external suspension) were excluded, and a total of 185 patients were enrolled in the final cohort. Based on the primary outcome set for the model, 49 patients (26.5%) were allocated to the PMV group. The medium age for the whole group was 0.7 years old (IQR,

0.39–1.54), with 44.0% of the patients being female. Regarding different CHD procedures, the STAT category was adopted for the risk stratification and differences were mainly observed in STAT 1, STAT 3, STAT 4, and STAT 5 categories. 22 patients (59%) in the PMV group were comorbid with tracheobronchomalacia. According to the grouping, the perioperative outcomes, including CPB, ACC, ICU stay, postoperative length of stay, reintubation, ECMO implication, postoperative tracheotomy, ventilator-associated pneumonia, pneumothorax, and in-hospital death were significantly different between the two groups ( $P < 0.05$ ). There were ten in-hospital deaths, eight of which belonged to the PMV group. Detailed information of baseline characteristics of the whole cohort was shown in Table 1 and Supplementary Table 1.

**Table 1** Baseline characteristics

	Overall (N = 185)	Mechanical ventilation duration		P-value
		≤ 2 weeks (N = 136)	> 2 weeks (N = 49)	
Gestational age, weeks	39.00 (38.00, 40.00)	39.00 (38.00, 40.00)	38.00 (37.00, 40.00)	0.4
Weight at birth, kg	3.10 (2.70, 3.50)	3.15 (2.70, 3.50)	3.10 (2.80, 3.40)	0.7
Age at CHD surgery, years	0.70 (0.39, 1.54)	0.74 (0.44, 1.73)	0.58 (0.25, 0.93)	0.004
Weight at CHD surgery, kg	7.2 (5.5, 10.0)	7.5 (6.0, 10.7)	6.1 (5.0, 7.7)	< 0.001
Height at CHD surgery, cm	66 (61, 78)	70 (63, 80)	63 (57, 69)	< 0.001
CPB duration, min	92 (56, 133)	72 (38, 115)	126 (96, 182)	< 0.001
ACC duration, min	48 (6, 80)	42 (0, 70)	77 (52, 114)	< 0.001
Mechanical ventilation, hours	36 (7, 375)	20 (5, 52)	777 (500, 1,323)	< 0.001
Postoperative ICU-stay, days	6 (2, 26)	4 (2, 8)	45 (33, 66)	< 0.001
Postoperative hospital-stay, days	20 (13, 40)	15 (11, 20)	53 (41, 82)	< 0.001
Gender				0.6
Male	104 (56%)	75 (55%)	29 (59%)	
Female	81 (44%)	61 (45%)	20 (41%)	
STAT category				< 0.001
STAT1	73 (39%)	67 (49%)	6 (12%)	
STAT2	55 (30%)	39 (29%)	16 (33%)	
STAT3	36 (19%)	21 (15%)	15 (31%)	
STAT4	16 (8.6%)	9 (6.6%)	7 (14%)	
STAT5	5 (2.7%)	0 (0%)	5 (10%)	
Complex CHD	119 (64%)	77 (57%)	42 (86%)	< 0.001
Tracheobronchomalacia	56 (30%)	27 (20%)	29 (59%)	< 0.001
Tracheobronchial compression	57 (31%)	46 (34%)	11 (22%)	0.14
Congenital tracheal stenosis	18 (9.7%)	17 (12%)	1 (2.0%)	0.046
Stenotic segment				0.057
Tracheal	74 (40%)	60 (44%)	14 (29%)	
Bronchus	111 (60%)	76 (56%)	35 (71%)	
Reintubation	26 (14%)	9 (6.6%)	17 (35%)	< 0.001
ECMO use	11 (5.9%)	0 (0%)	11 (22%)	< 0.001
Postoperative tracheotomy	25 (14%)	2 (1.5%)	23 (47%)	< 0.001
Ventilator-associated pneumonia	40 (22%)	10 (7.4%)	30 (61%)	< 0.001
Pneumothorax	8 (4.3%)	2 (1.5%)	6 (12%)	0.005
In-hospital deaths	10 (5.4%)	2 (1.5%)	8 (16%)	< 0.001

Value are presented with median (IQR) for continuous variables, number (percentage) for categorical variables. CHD: Congenital heart disease; CPB: Cardiopulmonary bypass; ACC: Aortic cross-clamping; ICU: Intensive care unit; ECMO: Extracorporeal membrane oxygenation. Complex CHD excludes simple shunts (atrial septal defect, ventricular septal defect, patent ductus arteriosus), vascular rings, and pulmonary artery slings.

**Data imputation**

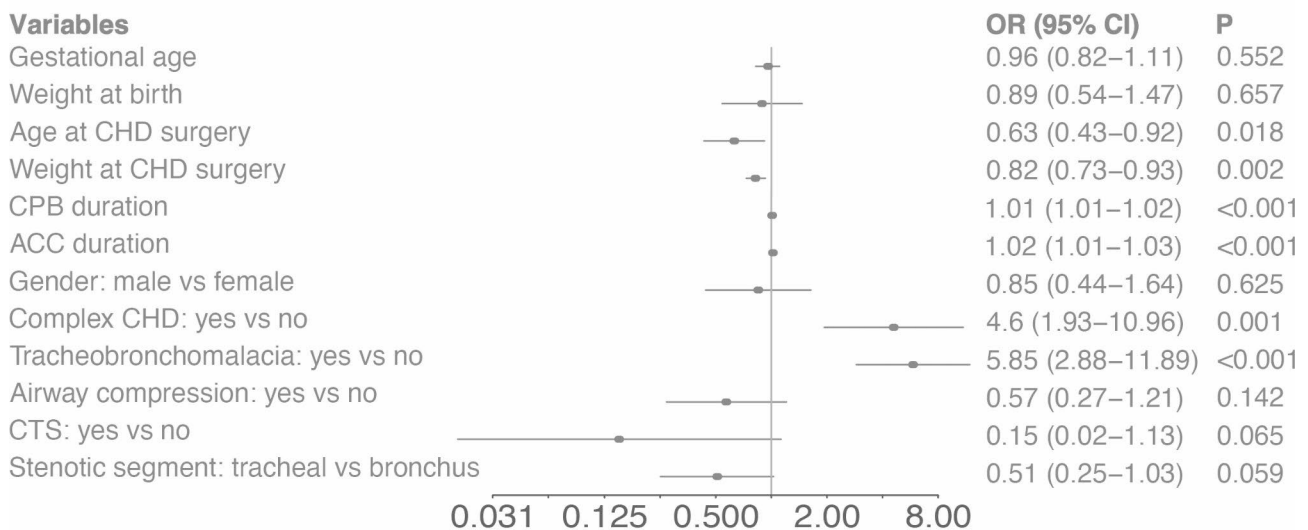
Several variables in the original dataset contained various degrees of missing data, including gestational age (18/185, 9.7%), birth weight (21/185, 11.4%), weight at CHD surgery (1/185, 0.5%), height at CHD surgery (4/185, 2.2%), cardiopulmonary bypass (CPB) duration (1/185, 0.5%) and aortic cross clamp (ACC) duration (1/185, 0.5%). Multiple imputation methods were utilized to fill in the missing values. The imputed dataset was compared with the original dataset, and the results showed that the imputation did not cause any significant bias (Supplementary Table 2).

**Predictive model**

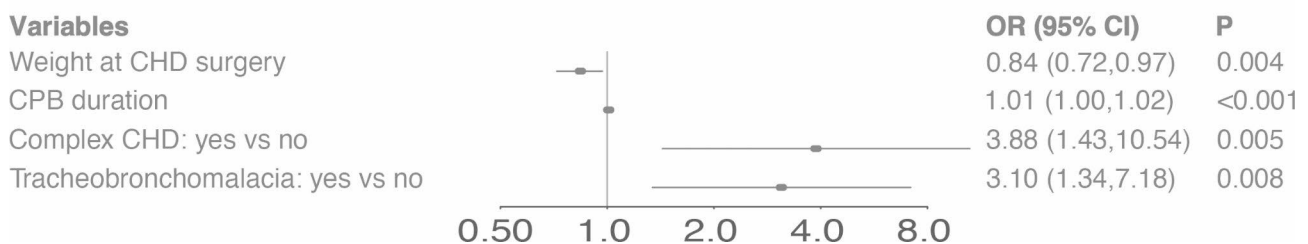
Twelve variables were included in the predictive model with univariate logistics regression analysis (Fig. 1A). Multivariate logistic regression indicated four independent risk factors for PMV, including weight at CHD surgery, CPB duration, complex CHD and comorbid tracheobronchomalacia (Fig. 1B). Since both complex

CHD and CPB duration represented the complexity of the surgical procedure, we built additional two different logistic regression models (Model 2 [weight at CHD surgery+complex CHD+comorbid tracheobronchomalacia] and Model 3 [weight at CHD surgery+CPB duration+comorbid tracheobronchomalacia]) to explore the contribution of two variables. The likelihood ratio test for the regression model showed statistically significant differences between Model 1, Model 2 and Model 3 (Model 1 vs. Model 2:  $P < 0.001$ ; Model 1 vs. Model 3  $P = 0.005$ ). As the calibration plot indicated, the predicted probability overlapped with the actual probability in all three models, showing the consistency between regression models and actual observations (Fig. 2A-C). The Hosmer-Lemeshow test also showed no statistically significant difference between the predicted and actual values (Model 1:  $\chi^2 = 3.041$ ,  $P = 0.932$ ; Model 2:  $\chi^2 = 6.773$ ,  $P = 0.561$ ; Model 3:  $\chi^2 = 3.061$ ,  $P = 0.931$ ). According to the ROC analysis, the AUC for Model 1, Model 2 and Model 3 were 0.847 (95% CI: 0.786–0.908), 0.801

**A Univariate logistic regression**



**B Multivariate logistic regression**



**Fig. 1** Factors associated with PMV. **(A)** Univariate logistic regression. Factors with predictive potential were included in the univariate logistic regression, and the result was presented with a forest plot. **(B)** Multivariate logistic regression. Four factors were included in the predictive model and presented with a forest plot, including weight at CHD surgery, CPB duration, complex CHD, and tracheobronchomalacia.  $P < 0.05$  was considered to be statistically significant

(95% CI: 0.729–0.874), 0.831 (95% CI: 0.765–0.890), and the recommended cut-off values for each model were 0.221, 0.334 and 0.319, respectively (Fig. 2D–F). Next, we examined the clinical performance of these three regression models with decision curve analysis (Fig. 2G). Compared to the control line, Model 1, Model 2 and Model 3 resulted in high net benefit values in a wide range of threshold probabilities, especially Model 1.

After evaluating the models for prediction accuracy and clinical implication, Model 1 was selected as the final prediction model. All variables in Model 1 were included in the multicollinearity test, and no multicollinearity was observed (Fig. 2H). A nomogram based on logistic regression was plotted to visualize the predictive model (Fig. 3). The 3, 5, and 10-fold cross-validation and the bootstrap validation indicated that the predictive model has good accuracy and no obvious over-interpretation (Supplementary Table 3).

The values (or classifications) of these perioperative indicators were used to score each patient's risk and thus the probability of PMV was calculated. According to the cut-off value of the ROC curve, patients were divided into high-risk and low-risk groups. In addition to the duration of mechanical ventilation, other important postoperative factors differed between groups based on predictive models, such as post-operative length of stay, ICU duration, and in-hospital deaths (Table 2).

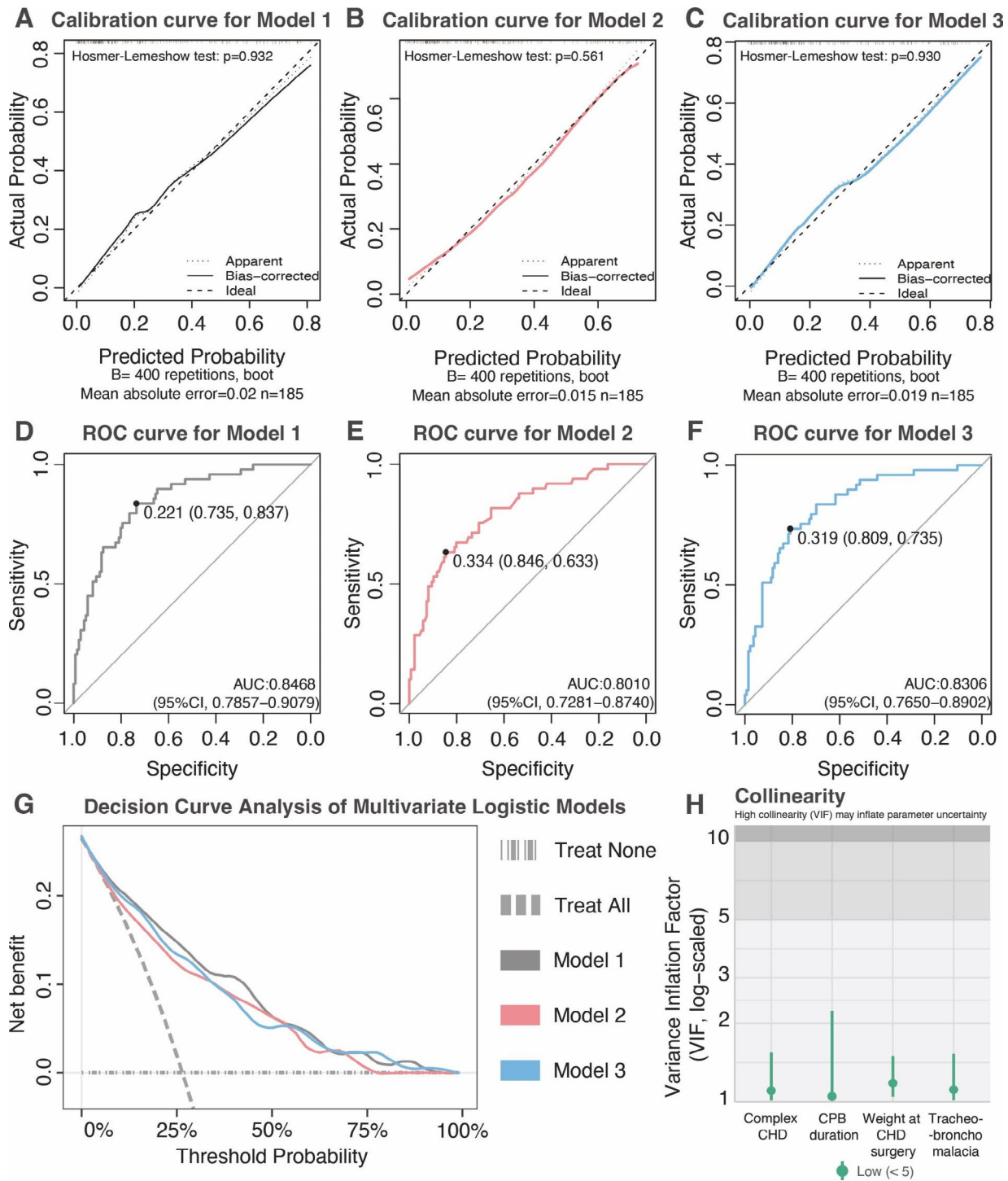
## Discussion

This is the first study focusing on CHD patients with AS who underwent conservative airway treatment. Plenty of studies have investigated the surgical outcomes of CHD patients who underwent airway surgery, while the data on AS without intervention are scarce. Riggs and colleagues retrospectively reviewed the associated airway anomaly in pediatric patients who underwent heart surgery from the STS-CHSD, and this study provided insights in patients with AS but without surgical intervention that the operative mortality of 5.9%, major morbidity of 21.2%, and postoperative tracheotomy of 5.6%, respectively [8]. As a result, we sought to take a deep insight into this population and provided more information on their perioperative characteristics, by retrospectively reviewing the patients who met the criteria from 2009 to 2022 in the National Center for Cardiovascular Center of China.

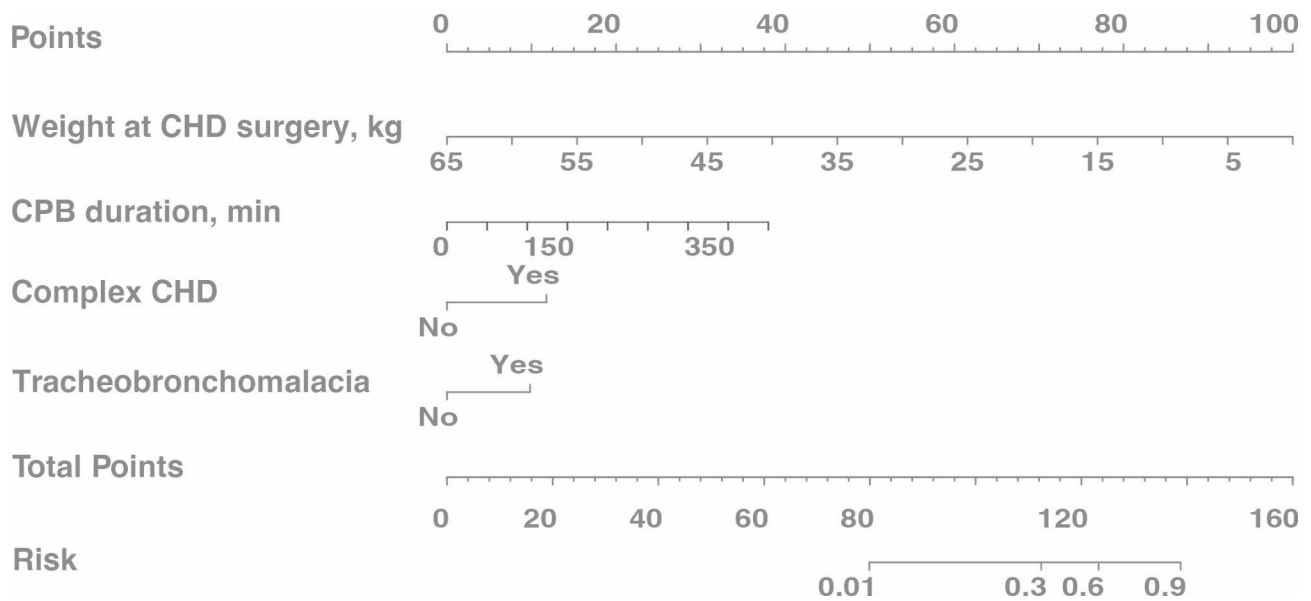
The mechanical ventilation duration is an important perioperative indicator for children undergoing cardiac surgery, especially children with airway anomalies, yet there was no consensus definition of PMV in children. According to a systemic review, this definition varied from 48 h to 6 months [15]. Polito et al. retrospectively investigated the mechanical ventilation duration of patients after complex congenital cardiac surgery, and

the cohort comprised 362 patients, of whom 41 (11%) required mechanical ventilation for  $\geq 7$  days (median ventilation duration for 362 patients: 1.5 days, range: 0–7 days) [13]. However, the mechanical ventilation duration of CHD patients was longer when complicated with airway anomaly. McMahon et al. reported that pediatric patients with CHD and bronchopulmonary dysplasia had a median postoperative mechanical ventilation duration of 15 days (range: 1–141) [16]. For patients with CHD and CTS, the median ventilation duration was 9 days (IQR, 5–20.75) [11]. Given that the coexisting airway stenosis may extend the mechanical ventilation duration compared to patients with CHD only, we established the standard of PMV at a higher level ( $\geq 14$  days). We also referenced results from PICU, where patients with various conditions, not limited to cardiovascular disease. A cohort enrolled children in PICU showed the incidence of PMV ( $\geq 14$  days) is 33.2%, identifying an elevated rate of extubation failure, increased hospitalization costs, and higher mortality after 1-month discharge in patients who received PMV [17].

The PMV is associated with ICU stay duration, re-intubation, perioperative complications, and increased mortality [18, 19]. There were several research focusing on the predictive factors for mechanical ventilation after pediatric cardiac surgery. Gaies et al. developed a model to predict the mechanical ventilation duration after pediatric cardiac surgery, revealing that age, prematurity, extracardiac/genetic anomalies, underweight, preoperative mechanical ventilation, higher STAT category (STAT 4 and 5), and cardiopulmonary bypass duration were the independent predictors [20]. According to a multicenter study performed by Gupta et al., the odds of mechanical ventilation after cardiac surgery were associated with patient characteristics, surgical risk category, and cardiac center volume [21]. Similarly, Maisat et al. identified risk factors for postoperative mechanical ventilation for pediatric pulmonary vein stenosis, including male sex, low body weight, preoperative oxygen supplement, high PVS severity score, intraoperative red blood cell transfusion low preintervention PaO<sub>2</sub>/FIO<sub>2</sub> ratio, and high preintervention right ventricular systolic pressure [22]. According to our study, four predictive factors were identified when focusing on children with CHD and AS: weight at CHD surgery, CPB duration, complex CHD, and comorbid tracheobronchomalacia. CHD, especially great vessel anomalies can cause airway compression, contributing to tracheobronchomalacia [23]. According to Chen et al., the combination of CHD and tracheobronchomalacia was associated with PMV, ICU-stay, hospital-stay, and mortality [24]. For a subset of patients with airway compression or tracheobronchomalacia, invasive airway intervention may be waived after the relief of compression or stenosis by cardiac surgery.



**Fig. 2** The diagnosis of the predictive model. **(A-C)** Calibration curves for logistic regression models. The predicted probability of PMV was presented on the x-axis, and the actual probability was presented on the y-axis. All three models passed the Hosmer-Lemeshow test ( $P > 0.05$ ); **(D-F)** ROC curves for logistic regression models. The ROC curves and optimal cut-off values for Model 1 (grey), Model 2 (red), and Model 3 (blue) were plotted; **(G)** Decision curve analysis for PMV prediction. We preferred the prediction model with higher net benefit values over a larger threshold probability range. All three prediction models had higher net benefits than the control line (treat all and treat none) over the full range of threshold probabilities **(H)** Diagnosis of multicollinearity of Model 1. The result showed no multicollinearity (Variance Inflation Factor < 5). Variables: Model 1: weight at CHD surgery, CPB duration, complex CHD and comorbid tracheobronchomalacia; Model 2: weight at CHD surgery, complex CHD and comorbid tracheobronchomalacia; Model 3: weight at CHD surgery, CPB duration and comorbid tracheobronchomalacia



**Fig. 3** The nomogram for predicting the risk of PMV. The corresponding total score was calculated based on the value of each variable. The corresponding risk of PMV was based on the total score

**Table 2** Postoperative features of low-/high-risk groups

Variables	Low-risk group (N=108)	High-risk group (N=77)	P-value
Mechanical ventilation, hours	16 (5, 36)	375 (70, 776)	< 0.001***
Postoperative ICU-stay, days	3 (1, 6)	24 (11, 46)	< 0.001***
Postoperative hospital-stay, days	14 (10, 20)	35 (20, 56)	< 0.001***
Reintubation	6 (5.6%)	20 (26%)	< 0.001***
ECMO use	1 (0.9%)	10 (13%)	< 0.001***
Postoperative tracheotomy	7 (6.5%)	18 (23%)	< 0.001***
Ventilator-associated pneumonia	11 (10%)	29 (38%)	< 0.001***
Pneumothorax	5 (4.6%)	3 (3.9%)	0.9
In-hospital deaths	1 (0.9%)	9 (12%)	0.002**

Values are presented with median (IQR) for continuous variables, number (percentage) for categorical variables. ICU: Intensive care unit; ECMO: Extracorporeal membrane oxygenation.

Patients included in our cohort had a relatively young age (median 0.70, IQR 0.39–1.54), which could be attributed to the growth of children’s airways. As their bodies grow, the length, diameter, and cross-sectional area of the airway increase, and this growth process continues into adulthood [25]. In comparison to the trachea in adults, the size of the trachea during infancy is approximately 50%, 36%, and 15% of the length, diameter, and cross-sectional area, respectively [26]. The growth of the trachea has been observed in patients with CTS as well, with the diameter approaching normal values by the age of nine [7].

Our study developed a nomogram-based prediction model and the corresponding risk scores could be calculated. The predictive model identified patients who had a high risk for PMV, and those patients may tend to

have worse postoperative outcomes, including postoperative ICU-stay and hospital-stay, reintubation, ECMO use, postoperative tracheotomy, and in-hospital death (Table 2). Therefore, early identification of this group is essential that more attention should be paid on intensive care of these patients in the perioperative period, and more comprehensive as well as routine surveillance of these patients should be set up after discharge to further improve the prognosis.

Efforts were made to shorten the mechanical ventilation duration. According to a randomized clinical trial by Blackwood et al., compared to usual care in the pediatric ICU, a sedation and ventilator liberation protocol including assessment of sedation levels, spontaneous breathing trials, and non-invasive ventilator resulted in a statistically significant reduction in time to the first successful extubation [27]. Tracheotomy during ventilator adoption is also considered to be beneficial in reducing mechanical ventilation. Each pediatric intensive care unit has different options for the timing of tracheostomy [28]. A recent review suggested that performing a tracheotomy early may improve important medical outcomes [29]. Other management included specific body positions for receiving mechanical ventilation and intraoperative protective ventilation [30, 31]. Strategies for reducing the duration of mechanical ventilation in patients with congenital heart disease combined with AS are still under discussion. However, one thing is certain, early identification of those at risk for PMV is necessary.

### Limitations

This study has several limitations to be addressed. First, as the present study was a single-center retrospective study, validation of an external cohort was lacking. The model's predictive power should be tested in an external cohort by calculating each patient's risk score and comparing it to their actual outcomes. Although we performed adequate internal validation to assess whether the model was overfitted, we were still unable to confirm whether the model was applicable to all patients. A subsequent prospective study may serve as an external validation cohort to evaluate the external applicability of this model. Second, patients included in the study suffered not only from AS but also various cardiac malformations, the influence of cardiac surgery could not be ignored, even if we tried to reduce this imbalance, the final results might still be influenced. Finally, our study included all the patients who met the inclusion criteria between July 2009 and January 2022, it is possible that the sample size was inadequate, albeit to the rarity of CHD complicated with AS [32]. The optimal sample size for the predictive model was calculated according to the 4-step sample size calculation method proposed by Riley et al. [32]. Since no similar prediction model was available for our reference, the parameters we set when using the 4-step method were based on the model we built, the calculation results could only be used for model evaluation and subsequent model refinement. With an outcome incidence of 0.265, an AUC value of 0.847, and 4 variables planned to be included as candidates, a sample size of 300 was calculated using the *pmsampsize* package of R, and 80 of these outcome events should be observed.

### Conclusions

We provided a predictive nomogram model to predict the postoperative PMV in patients with CHD and AS who underwent non-surgical airway intervention. The nomogram we plotted could be used to identify those patients at risk. These patients might be benefited from early identification, with more intensive monitoring and extra airway management.

#### List of abbreviations

ACC	Aortic cross clamp
AIC	Akaike information criterion
AS	Airway stenosis
AUC	Area under curve
CHD	Congenital heart diseases
CPB	Cardiopulmonary bypass
CT	Computerized tomography
CTS	Congenital tracheal stenosis
DCA	Decision curve analysis
ECMO	Extracorporeal membranous oxygenation
ICU	Intensive care unit
IQR	Interquartile range
MDT	Multidisciplinary treatment
ROC	Receiver operating characteristic

STAT	Society of Thoracic Surgeons- European Association for Cardiothoracic Surgery
STS-CHSD	Society of Thoracic Surgeons Congenital Heart Surgery Database
VIF	Variance inflation factor

### Supplementary Information

The online version contains supplementary material available at <https://doi.org/10.1186/s12887-023-04160-5>.

Supplementary Material 1

Supplementary Material 2

Supplementary Material 3

### Acknowledgements

Not applicable.

### Authors' contributions

Qiyu He and Yuze Liu: Conceptualization, data processing, manuscript writing. Zheng Dou and Kai Ma: Investigation. Shoujun Li: Supervision. All authors approved the final manuscript as submitted and agree to be accountable for all aspects of the work.

### Funding

This study was supported by the CAMS Innovation Fund for Medical Sciences (CIFMS, 2020-I2M-C&T-A-009), Capital Health Research and Development of Special Fund (2022-1-4032), and the National High Level Hospital Clinical Research Funding (2022-GSP-GG-19).

### Data Availability

The data used in this current study are available from the corresponding author on reasonable request.

### Declarations

#### Competing interests

The authors declare no competing interests.

#### Ethical approval

This study was approved by the ethical review board of Fuwai Hospital (NO. 2021 – 1506). Informed consents were obtained from a parent and/or legal guardian who understood and participated in the study. All methods were carried out in accordance with relevant guidelines and regulations. All procedures performed in this study are in accordance with the Declaration of Helsinki.

#### Consent for publication

Not applicable.

Received: 15 May 2023 / Accepted: 26 June 2023

Published online: 12 July 2023

### References

- Lee YS, Jeng MJ, Tsao PC, Soong WJ, Chou P. Prognosis and risk factors for congenital Airway Anomalies in children with congenital heart disease: a Nationwide Population-Based study in Taiwan. *PLoS ONE*. 2015;10:e0137437.
- Sulkowski JP, Deans KJ, Asti L, Mattei P, Minneci PC. Using the Pediatric Health Information System to study rare congenital pediatric surgical diseases: development of a cohort of esophageal atresia patients. *J Pediatr Surg*. 2013;48:1850–5.
- Guillemaud JP, El-Hakim H, Richards S, Chauhan N. Airway pathologic abnormalities in symptomatic children with congenital cardiac and vascular disease. *Arch Otolaryngol Head Neck Surg*. 2007;133:672–6.



4. Herrera P, Caldarone C, Forte V, Campisi P, Holtby H, Chait P, et al. The current state of congenital tracheal stenosis. *Pediatr Surg Int*. 2007;23:1033–44.
5. Hofferberth SC, Watters K, Rahbar R, Fynn-Thompson F. Manage Congenital Tracheal Stenosis *Pediatr*. 2015;136:e660–669.
6. Sengupta A, Murthy RA. Congenital tracheal stenosis & associated cardiac anomalies: operative management & techniques. *J Thorac Dis*. 2020;12:1184–93.
7. Cheng W, Manson DE, Forte V, Ein SH, MacLusky I, Papsin BC, et al. The role of conservative management in congenital tracheal stenosis: an evidence-based long-term follow-up study. *J Pediatr Surg*. 2006;41:1203–7.
8. Riggs KW, Zafar F, Jacobs ML, Jacobs JP, Thibault D, Guleserian KJ, et al. Tracheal surgery for airway anomalies associated with increased mortality in pediatric patients undergoing heart surgery: society of thoracic Surgeons database analysis. *J Thorac Cardiovasc Surg*. 2021;161:1112–1121e1117.
9. Arcieri L, Pak V, Poli V, Baggi R, Serio P, Assanta N, et al. Tracheal surgery in children: outcome of a 12-year survey. *Interact Cardiovasc Thorac Surg*. 2018;26:660–6.
10. Chen L, Zhu L, Wang H, Lu Z, Xu Z, Du X, et al. Surgical management strategy of slide tracheoplasty for infants with congenital tracheal stenosis. *J Thorac Cardiovasc Surg*. 2022;163:2218–28.
11. Ramaswamy M, Yeh YT, Varman R, McIntosh N, McIntyre D, Fedevych O, et al. Staging of Surgical Procedures in Comorbid congenital tracheal stenosis and congenital Cardiovascular Disease. *Ann Thorac Surg*. 2020;109:1889–96.
12. Székely A, Sápi E, Király L, Szatmári A, Dinya E. Intraoperative and postoperative risk factors for prolonged mechanical ventilation after pediatric cardiac surgery. *Paediatr Anaesth*. 2006;16:1166–75.
13. Polito A, Patorno E, Costello JM, Salvin JW, Emami SM, Rajagopal S, et al. Perioperative factors associated with prolonged mechanical ventilation after complex congenital heart surgery. *Pediatr Crit Care Med*. 2011;12:e122–126.
14. Chauhan JC, Slamon NB. The impact of multiple viral respiratory infections on outcomes for critically ill Children. *Pediatr Crit Care Med*. 2017;18:e333–8.
15. Colletti J Jr, Azevedo RT, de Oliveira Caino FR, de Araujo OR. Prolonged mechanical ventilation in children: review of the definition. *Pediatr Crit Care Med*. 2021;22:e588–93.
16. McMahon CJ, Penny DJ, Nelson DP, Ades AM, Al Maskary S, Speer M, et al. Preterm infants with congenital heart disease and bronchopulmonary dysplasia: postoperative course and outcome after cardiac surgery. *Pediatrics*. 2005;116:423–30.
17. Liu Y, Wang Q, Hu J, Zhou F, Liu C, Li J, et al. Characteristics and risk factors of children requiring prolonged mechanical ventilation vs. non-prolonged mechanical ventilation in the PICU: a prospective single-center study. *Front Pediatr*. 2022;10:830075.
18. Harris KC, Holowachuk S, Pitfield S, Sanatani S, Froese N, Potts JE, et al. Should early extubation be the goal for children after congenital cardiac surgery? *J Thorac Cardiovasc Surg*. 2014;148:2642–7.
19. Gupta S, Boville BM, Blanton R, Lukasiewicz G, Wincek J, Bai C, et al. A multicentered prospective analysis of diagnosis, risk factors, and outcomes associated with pediatric ventilator-associated pneumonia. *Pediatr Crit Care Med*. 2015;16:e65–73.
20. Gaies M, Werho DK, Zhang W, Donohue JE, Tabbutt S, Ghanayem NS, et al. Duration of postoperative mechanical ventilation as a Quality Metric for Pediatric Cardiac Surgical Programs. *Ann Thorac Surg*. 2018;105:615–21.
21. Gupta P, Rettiganti M, Gossett JM, Yeh JC, Jeffries HE, Rice TB, et al. Risk factors for mechanical ventilation and reintubation after pediatric heart surgery. *J Thorac Cardiovasc Surg*. 2016;151:451–8. e453.
22. Maisat W, Yuki K. Predictive factors for postoperative intensive care unit admission and mechanical Ventilation after Cardiac catheterization for Pediatric Pulmonary Vein Stenosis. *J Cardiothorac Vasc Anesth*. 2022;36:2500–8.
23. Svetanoff WJ, Zendejas B, Smithers CJ, Prabhu SP, Baird CW, Jennings RW, et al. Great vessel anomalies and their impact on the surgical treatment of tracheobronchomalacia. *J Pediatr Surg*. 2020;55:1302–8.
24. Chen Q, Langton-Hewer S, Marriage S, Hayes A, Caputo M, Pawade A, et al. Influence of tracheobronchomalacia on outcome of surgery in children with congenital heart disease and its management. *Ann Thorac Surg*. 2009;88:1970–4.
25. Griscom NT, Wohl ME. Dimensions of the growing trachea related to age and gender. *AJR Am J Roentgenol*. 1986;146:233–7.
26. Di Cicco M, Kantar A, Masini B, Nuzzi G, Ragazzo V, Peroni D. Structural and functional development in airways throughout childhood: children are not small adults. *Pediatr Pulmonol*. 2021;56:240–51.
27. Blackwood B, Tume LN, Morris KP, Clarke M, McDowell C, Hemming K, et al. Effect of a sedation and Ventilator Liberation Protocol vs Usual Care on Duration of Invasive Mechanical Ventilation in Pediatric Intensive Care units: a Randomized Clinical Trial. *JAMA*. 2021;326:401–10.
28. Wakeham MK, Kuhn EM, Lee KJ, McCrory MC, Scanlon MC. Use of tracheostomy in the PICU among patients requiring prolonged mechanical ventilation. *Intensive Care Med*. 2014;40:863–70.
29. Abdelaal Ahmed Mahmoud MAA, Younis M, Jamshidi N, Hussein HA, Farag E, Hamza MK, et al. Timing of Tracheostomy in Pediatric Patients: a systematic review and Meta-analysis. *Crit Care Med*. 2020;48:233–40.
30. Rivas-Fernandez M, Roque IFM, Diez-Izquierdo A, Escribano J, Balaguer A. Infant position in neonates receiving mechanical ventilation. *Cochrane Database Syst Rev*. 2016;11:CD003668.
31. Ladha K, Vidal Melo MF, McLean DJ, Wanderer JP, Grabitz SD, Kurth T, et al. Intraoperative protective mechanical ventilation and risk of postoperative respiratory complications: hospital based registry study. *BMJ*. 2015;351:h3646.
32. Riley RD, Ensor J, Snell KIE, Harrell FE Jr, Martin GP, Reitsma JB, et al. Calculating the sample size required for developing a clinical prediction model. *BMJ*. 2020;368:m441.

## Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.