



Reduce Mortality in Robin Sequence Patient with Cardiac Anomalies

Yunyan Z^{1*}, Xiaohao L¹, Yang L², Lijuan L³, Yingqiu C¹ and Youqiang L⁴

¹Department of Stomatology, Guangzhou Women and Children's Medical Center, China

²Guangzhou University of Chinese Medicine, China

³Department of Heart Center, Guangzhou Women and Children's Medical Center, China

⁴Department of Laboratory Medicine, The Affiliated Hexian Memorial Hospital of Southern Medical University, China

Abstract

Cardiac anomalies are frequently encountered in Robin Sequence (RS) neonates with upper airway obstruction. The RS patients were dyspnea, fed difficulties, with cardiac abnormalities caused by cardiac dysfunction, aggravating hypoxia, poor nutrition, increased surgical risk. Mortality rate of RS patients with cardiac anomalies was high. Neonatal distraction (Mandibular Distraction Osteogenesis, MDO) in severe micrognathia patients have been shown to be effective method in treating upper airway obstruction. In our hospital, MDO and cardiac surgery were done at the same time, upper airway obstruction and cardiac structural abnormalities due to hypoxia, can simultaneously treated, no one died, RS patients gain the most weight and benefits. It's a safe and effective way to do both surgeries at the same time for the RS patient with cardiac anomalies.

Keywords: Robin sequence; Cardiac anomalies; Surgery reduce mortality

Introduction

Robin Sequence (RS) was first described by the French Stomatologist Pierre Robin in 1923 and is characterized by the triad of micrognathia, subsequently leading to glossoptosis and varying degrees of upper airway obstruction [1]. RS is a congenital condition occurring in approximately 1 in 5600-8000 live births [2,3]. Recently, an international consensus was achieved regarding the three distinguishing characteristics (micrognathia, glossoptosis, and upper airway obstruction) that should be included in the diagnosis of RS in newborns. Cardiac anomalies and cleft palate and is frequently encountered [4].

The incidence of cardiac anomalies in Robin sequence has been previously reported at 7% to 22% [5-8]. Cardiac anomalies include ventricular septal defect, patent foramen ovale, patent ductus arteriosus, pulmonary hypertension. In the study by [5], 30% of patients who died had cardiac anomalies. Monroe and Ogo [7] reported a 21.5% incidence of congenital heart disease in 65 patients with Robin syndrome [7]. However, mortality rate of PS patient with cardiac anomaly was 39.3% [9]. Congenital heart disease was the important factor for mortality in RS patients.

However how to reduce mortality in RS patients with severe structural abnormalities of the heart. Mandibular Distraction Osteogenesis (MDO) has been shown to be successful in treating upper airway obstruction caused by micrognathia in pediatric RS patients; however, high operation risk exits [10]. The objective of this study was to given the improvements to reduce the mortality in patient with RS and congenital heart disease.

Patient and Methods

All children with Robin sequence admitted to the neonatal intensive care unit (<28 d) from January of 2017 to December of 2019 were identified. Inclusion criteria were children diagnosed with RS (micrognathia, glossoptosis, and upper airway obstruction, with or without cleft palate) and cardiac anomalies (except patent foramen ovale). Admitted with a small mandible and no evidence of respiratory obstruction were excluded. Any evidence of respiratory compromise in the presence of a small jaw requiring at least supplemental oxygen or positioning changes was considered to be sufficient for a diagnosis of RS. In contrast, a child with a small jaw and no breathing problems (not requiring supplemental oxygen, positioning changes, or other interventions) was not considered to

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*Correspondence:

Yunyan Zhang, Department of Stomatology, Guangzhou Women and Children's Medical Center, Guangzhou Medical University, Guangzhou, China, E-mail: yunyangirl@126.com

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Table 1: Demographics and RS with cardiac malformation characteristics.

	Sex	Gestational weeks	Weight	G	P	Cardiac anomalies			
						ASD	VSD	PDA	PHA
Patient 1	female	38	3	2	2	-	-	-	-
Patient 2	female	40	3	1	1	-	-	-	-
Patient 3	male	40	3.5	4	3	-	-	-	-
Patient 4	female	40	3.3	2	1	-	-	-	-
Patient 5	male	40	3.2	4	2	-	-	-	-
Patient 6	male	37	3.05	4	2	-	-	-	-
Patient 7	male	40	2.5	1	1	-	-	-	-
Patient 8	male	38	3.5	3	3	-	-	-	-
Patient 9	male	37	3.2	6	2	-	-	-	-
Patient 10	male	40	3.02	1	1	-	-	-	-

G: Gestation; P: Parturition; ASD: Atrial Septal Defect; VSD: Ventricular Septal Defect; PDA: Patent Ductus Arteriosus; PHA: Pulmonary Arterial Hypertension

Table 2: Hospitalization time and ventilator use time.

	Patient	Hospitalization time (day)	Ventilator use time (day)
MDO and cardiac surgery were done at separate time	Patient 10	102	19
	Patient 3	61	9
	Patient 4	64	24
	Patient 6	22	4
	Patient 5	Give up the treatment	
MDO and cardiac surgery were done at the same time	Patient 7	37	5
	Patient 8	26	8
	Patient 9	19	3
	Patient 1	21	9
	Patient 2	32	5

Table 3: Weight gain in two patients.

	Sex	First surgery			Second surgery			Third surgery		
		Operation	Age (day)	Weight (kg)	Operation	Age (day)	Weight (kg)	Operation	Age (day)	Weight (kg)
Separate group	Male	Cardiac surgery	17	3.5	MDO	176	3.5	Removal of mandibular distractor	377	9.2
Same time group	Male	Cardiac surgery + MDO	21	2.92	Removal of mandibular distractor	180	8	Palatoplasty	365	12

have RS. Guangzhou women and children's medical center Review Board approval was obtained before the start of the study.

Patient and Indications for Surgery

Virtually ten patients with III grade of RS [11] and cardiac anomalies were in this study. Three patients underwent cardiac surgery, were discharged from hospital and MDO after three months of recuperation. One patient underwent cardiac surgery 20 days later, followed by MDO. One patient underwent cardiac surgery, three days later, was given up treatment. Five patients underwent cardiac surgery and mandibular distraction at the same day, cardiac surgery first, mandibular distraction second. The patients with PDA were treated by thoracotomy through median thoracotomy. Repair of foramen ovale and ventricular septal defect by cardiopulmonary bypass and thoracotomy. Twenty III grade RS patients without cardiac anomalies were included.

Statistical Analysis

Descriptive statistics are presented as mean \pm SD and frequency

(percentage) for continuous and categorical variables, respectively.

Outcome Measures

Identification

Identifications of the patient were as in Table 1. The birth weight, gestational weeks and gender of RS patient were similar with or without cardiac malformations, occur to males. And the number of gestation (2.7 ± 1.6) and parturitions (1.6 ± 0.91) was significantly higher than that of the children without cardiac malformations, gestation (1.7 ± 0.5), parturitions (1.3 ± 0.3). Pulmonary arterial hypertension was found in all children with cardiac malformation.

Clinical outcome

Hospitalization time and ventilator use time in RS patients without cardiac malformation (33.36 ± 18.71), (7.18 ± 1.47) was similar as in group of MDO and cardiac surgery at the same time, (27 ± 6.7), (6 ± 2.19), respectively.

Hospitalization time and ventilator use time for separate

operations was significantly longer than that for both operations. Patient 10 was hospitalized for 102 days, which was related to the duration of non-invasive ventilation and postural therapy. Patient 4 was difficulty in evacuation of breath machine after heart operation for 20 days, then MDO was operated, the breath machine was withdrawn smoothly after 4 days. Average ventilator-carried time was 6.8 days in simultaneous operation group; it is 17.3 days in the other group. After matching the two groups of patients, two patients were select; children underwent both operations at the same time gained more weight.

Discussion

Cardiac anomalies is a difficult problem in neonates with RS, mortality rate is high. The RS patients were dyspnea, feeding difficulties, with cardiac abnormalities caused by cardiac dysfunction, aggravating hypoxia, poor nutrition, increased surgical risk. It is difficult to make the decision which surgery was first. Cardiac surgery was performed first, prolonged ventilatory assist time; even it makes difficulties to evacuate the ventilator. However, if MDO first, the patient is at high risk, because of the unstable circulation. In our hospital, cardiac surgery and MDO were performed in the same day, cardiac surgery first, MDO second, average ventilator-carried time and hospitalization time was shorten, and the patient gained more weight.

Mortality rate was high in children untreated PAH [12]. In RS patient, VSD with left ventricular to right atrial shunt, blood enters the right ventricle, increased blood to the lungs leads to increased pulmonary arterial pressure. The ratio of right to left shunt in patients with PAH is not low. The foramen ovale were reopened by most of the intracardiac shunts, and PDA aggravating hypoxia, hypoxia causes pulmonary artery contraction, influence pulmonary gas exchange aggravates PAH and hypoxia [13]. In healthy people exposed to hypoxia, the potential arteriovenous branch in the lungs can be reopened. Therefore, it can be alleviates PAH to compensate for pulmonary vascular resistance, but aggravated hypoxia. RS patient suffered from hypoxia due to upper airway obstruction, potential arteriovenous branch maybe reopened, aggravated hypoxia. 71.4% of RS patients who died had cardiac disease [14]. Therefore, attention should be paid to PAH during the treatment.

So breaking hypoxia is the key to treat RS patients with cardiac malformation. In our department, nutritional support and oxygen inhalation were given several days before operation, improves internal environment. Cardiac surgery and MDO operated were performed on the same day, upper airway obstruction and cardiac structural abnormalities due to hypoxia, can simultaneously treated, RS patients gain the more weight, shorten the mechanical ventilation period as well as the hospital length of stay.

It is a safe and feasible way for neonatal RS patients to do cardiac surgery and MDO on the same day. In the future, we will study this in a larger sample to evaluate safety and explore complications.

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