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Pulse Oxygen Saturation Measured in Standing and Squatting Positions May Be Useful in Evaluating Tetralogy of Fallot

Qiang Li,[1,2,*] Jinzhou Zhang,[1,*] Baopin Li,[2] Wen Wang,[3] Jincheng Liu,[1] Hailong Zhu,[1] Hongbing Wang,[1] Shiqiang Yu,[1] Qin Cui,[1] Guocheng Sun,[1] Xiaochen Wu,[1] Dinghua Yi[1]

[1]Department of Cardiovascular Surgery and [3]Department of Traditional Chinese Medicine, Xijing Hospital, the Fourth Military Medical University, Xi'an 710032, People's Republic of China; [2]Department of Cardiothoracic Surgery, the Second Affiliated Hospital, the Shanxi Medical University

ABSTRACT

Background: Left-ventricular end-diastolic volume index (LVEDVI) and Nakata index, which in most cases are evaluated by echocardiography and cardiac catheterization, are 2 important predictors for the prognosis of surgical correction of tetralogy of Fallot (TOF). Nonetheless, performing these procedures on TOF patients is not always feasible. We therefore investigated whether the difference in transcutaneous pulse oxygen saturation between the standing position and squatting position (ΔSPO2) reflected the LVEDVI and Nakata index, allowing ΔSPO2 to be used as a supplement to echocardiography and cardiac catheter evaluation.

Methods: Nineteen TOF patients (11 boys, 8 girls; median age 6 years) were randomly selected for this study. In each patient, we used a pulse oximeter placed on the left index finger to measure transcutaneous pulse oxygen saturation, first with the patient in a standing position and then with the patient in a squatting position. We then performed correlational analyses of ΔSPO2 and the LVEDVI or Nakata index.

Results: The mean SPO2 was 79% ± 4% in standing patients and 84% ± 4% in squatting patients, and the mean ΔSPO2 was 5% ± 3%. The LVEDVI was 41 ± 5 mL/m^2, and the Nakata index was 188 ± 37 mm^2/m^2. ΔSPO2 correlated with both LVEDVI ($\gamma = 0.854$, $P < .05$) and the Nakata index ($\gamma = 0.641$, $P < .05$).

Conclusion: For patients with TOF, the ΔSPO2 between SPO2 measured in a standing and squatting position has a positive correlation with the LVEDVI and Nakata index. Thus these measurements may be used as supplemental factors in evaluating LV function and performing a preoperative assessment of the pulmonary artery.

Qiang Li and Jinzhou Zhang contributed equally to this work.

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Correspondence: Jinzhou Zhang, Department of Cardiovascular Surgery, Xijing Hospital, the Fourth Military Medical University, Xi'an 710032, People's Republic of China; 86-29-84775311; fax: 86-29-83210092 (e-mail: jinzhouzhang2006@yahoo.com) or Dinghua Yi, Department of Cardiovascular Surgery, Xijing Hospital, the Fourth Military Medical University, Xi'an 710032, People's Republic of China; 86-29-84775307; fax: 86-29-83210092 (e-mail: yidh@fmmu.edu.cn).

INTRODUCTION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease leading to cyanosis after the age of 1 year. Progressive hypoxia, cyanotic spells, cerebral infarction or abscess, and endocarditis are the major causes of morbidity and mortality from this disease [Horneffer 1990]. The first successful repair of TOF was performed by C. Walton Lillehei and his team in 1954 [Lillehei 1955]. Today, for a patient with "classic" pulmonary stenosis and without complicating associated anomalies such as absent pulmonary valve syndrome or complete artrial ventricular canal defects, TOF repair has a mortality risk of less than 2% [Karl 1992]. Some risk factors, however, continue to affect mortality and morbidity after surgical correction. For example, the status of the left ventricular (LV) and pulmonary arteries, most commonly evaluated by LV end-diastolic volume index (LVEDVI) and Nakata index, respectively, is the key factor in determining the outcome of TOF surgical repair [Ghai 2002].

Initially cardiologists and radiologists evaluated the LVEDVI and Nakata index by cardiac catheterization [Krabill 1987; Boxt 1994]. Despite the accuracy of this method, however, the use of this technique has been limited, especially in developing countries, because it is both invasive and demanding. Color Doppler echocardiography provided a new and noninvasive way to assess LVEDVI and the Nakata index. This method remains the primary diagnostic imaging tool but is not always available when planning surgical correction of TOF, because of a limited acoustic window and the inexperience of imagers. Therefore, we investigated the correlation between the difference in pulse-oxygen saturation measured in patients in standing and squatting positions (ΔSPO2), and the LVEDVI and Nakata index. We sought to find a new, accurate, and noninvasive supplement to color Doppler for evaluating the development of the LV and pulmonary artery in TOF patients.

MATERIALS AND METHODS

Study patients were 19 hospital patients and hospital-based outpatients with TOF with simple pulmonary stenosis diagnosed between January and May 2008. Patient age ranged from 2 to 14 years (median, 6 years). The chairperson of our

Table 1. Patient SPO2, LVEDVI, and Nakata Index*

Patient No.	SPO2, %			LVEDVI, mL/m^2	Nakata Index, mm^2/m^2
	Standing	Squatting	ΔSPO2		
1	78	81	3	37	156
2	81	88	7	41	190
3	80	86	6	39	183
4	77	79	2	34	163
5	76	86	10	47	230
6	83	85	2	36	241
7	81	84	3	41	203
8	79	81	2	37	158
9	77	83	6	49	160
10	75	86	11	49	167
11	76	83	7	45	186
12	71	76	5	40	155
13	80	88	8	47	259
14	77	82	5	40	141
15	79	83	4	37	160
16	85	89	4	41	205
17	86	88	2	36	167
18	85	90	5	39	265
19	81	87	6	41	184
Mean ± SD	79 ± 4	84 ± 4	5 ± 3	41 ± 5	188 ± 37

*SPO2 indicates pulse oxygen saturation; LVEDVI, left ventricular end-systolic volume index; ΔSPO2, difference of pulse oxygen saturation.

institutional review board approved the study. In addition, individual consent was obtained from each patient or their guardians before data collection and evaluation.

In this study, TOF was defined as described by Fallot [Neill 1994]. The 4 components of this malformation are: obstruction to right ventricular outflow, overriding of the aorta, ventricular-septal defect, and right ventricular hypertrophy. The criteria for exclusion of patients from the study were: (*a*) uncontrolled hypotension, (*b*) TOF with pulmonary atresia and absent pulmonary valve found during surgery, (*c*) other TOF-associated cardiac anomalies found during surgery, and (*d*) TOF unconfirmed during surgery.

Transcutaneous pulse oxygen saturation monitoring was carried out using a pulse oximeter (NPB-40; Nellcor, Boulder, CO, USA) with a skin sensor held in the left forefinger, first with the patient in a standing position and then in a squatting position. Room temperature was maintained in the range of 16°C-18°C, and the patients' arms were exposed. Skin temperature was in the range of 19°C-26°C. For 3 days before the test, patients received no inotropic agents or nonspecific β-blockers. Patients had not been given oxygen for at least 30 minutes before test. The data were used only if the skin sensor gave stable data 1 minute after the test.

All the patients underwent color Doppler examination performed by the same doctor using the same machine (Sonos 5500; Hewlett Packard, Palo Alto, CA, USA). Left and right pulmonary diameters were measured and Nakata indices were calculated according to formula reported by Nakata et al [1984]. Thus pulmonary artery size was the sum of the cross-sectional areas of the right and left pulmonary arteries, indexed to body surface area: (left pulmonary artery area + right pulmonary artery area)/(body surface area) (mm^2/m^2).

The LV long diameter, depth, transverse diameter, and LVEDVI were measured and obtained directly from 2-dimensional echocardiography. During physical examination, we also measured breathing frequency and cardiac frequency to determine if patients were in basal status.

We performed statistical analysis by using SPSS 14.0 for Windows. Numeric variables were expressed as mean ± SD. The Pearson's correlation test was used to analyze the variables. *P* values of 0.05 or less were considered statistically significant.

RESULTS

The transcutaneous pulse oxygen saturation measured while patients were in the standing position was 79% ± 4% and measured while patients were in the squatting position was 84 ± 4%. The ΔSPO2 between the different positions was 5% ± 3%. The LVEDVI was 41 ± 5 and the Nakata index was 188 ± 37 (Table 1).

We observed a significant and fairly strong positive correlation between LVEDVI and ΔSPO2 ($\gamma = 0.854$, $P < .05$). The coefficient of determination (γ^2) was 0.729. Likewise, similar positive correlations were found between the Nakata index and squatting SPO2 ($\gamma = 0.641$, $P < .05$) and between the difference in standing SPO2 and squatting SPO2 ($\gamma = 0.748$, $P < .05$) (Table 2).

DISCUSSION

In this study we found a fairly strong positive correlation between LVEDVI and ΔSPO2; indeed, approximately 73% of the ΔSPO2 variation could be explained by LVEDVI. In addition, we found that the Nakata index correlated with standing SPO2 and squatting SPO2. These findings demonstrated that both standing and squatting SPO2 were determined to a great extent by pulmonary resistance.

TOF occurs in approximately 1 in 3600 live births, is the cause of 5% to 7% of congenital heart lesions, and accounts for 3.5% of infants born with congenital heart disease. It is a progressive condition with an unfavorable natural history [Pozzi 2000]. Without surgical intervention, most patients die in childhood; survival rates are 66% at 1 year, 40% at 3 years, 11% at 20 years, and 3% at 40 years [Bertranou 1978]. Surgical correction of TOF has progressed significantly since the first repair was performed more than 60 years ago, and the efficiency of the surgery has improved [Lillehei 1986]. Currently, surgical repair of TOF can be accomplished with an actuarial survival rate of 98% [Knott-Craig 1998], even with the trend to perform repair surgery early in life [Giannopoulos 2002]. Many factors influence the outcome of total correction, such as age, pulmonary artery index, and LV developmental condition. Therefore, accurate evaluation of the pulmonary artery and LV developmental condition is crucial to the prognosis [Kirklin 1992].

LV function, often evaluated by LVEDVI, is usually accepted as a selective criterion for total surgical correction of TOF and is an independent risk factor for postoperative low cardiac output [Graham 1977; Nomoto 1984] and therefore useful in prognosis. With the further development of surgical techniques, perioperative support, and intraoperative myocardial preservation, the small LV as a contraindication to total correction is being reevaluated [Wu 1996]. The abovementioned advances, however, eliminate only some adverse effects and improve survival and extend indications for surgery only to a limited extent. The main obstacles to total surgical correction have not yet been overcome. Total correction be performed safely only in patients with an effective LV systolic stroke. A small LV will continue to be a contraindication in the near future, until evaluation can be improved.

Table 2. Pearson Correlations between LVEDVI, Standing SPO2, Squatting SPO2, ΔSPO2, and Nakata Index in Tetralogy of Fallot (n = 19)*

	LVEDVI	Standing SPO2	Squatting SPO2	ΔSPO2
Standing SPO2	−0.360			
Squatting SPO2	0.245	0.748		
ΔSPO2	0.854†	−0.402	0.307	
Nakata index	0.195	0.469	0.641†	0.213

*SPO2 indicates pulse oxygen saturation; LVEDVI, left ventricular end-systolic volume index; ΔSPO2, difference of pulse oxygen saturation.

†$P < .05$, 1-tailed.

The pulmonary artery index (Nakata index) is used to assess pulmonary artery width and the degree of capillary bed development. This measurement is useful in making treatment decisions, and developing surgical strategies for correction, and predicting postoperative outcome in TOF patients. The morphology and physiology of the pulmonary arteries are the 2 main factors in determining the Nakata index because the distribution and size of the pulmonary arteries and the efficiency of the pulmonary circulation are critical factors for successful surgery. In patients with TOF, the feasibility of complete operative repair is largely dependent on the underlying distribution and size of the pulmonary arteries [Potapov 2001]. Children with small Nakata index ratios are poor candidates for complete repair because of the inability of the small pulmonary arteries to handle the increased blood flow after complete repair [Sharma 1989].

Traditionally, the Nakata index and LVEDVI have been assessed by heart ventriculography or echocardiography. Angiography, which has been used for preoperative evaluation of the coronary arteries and peripheral pulmonary circulation, is the traditional standard criterion for evaluating pulmonary arterial morphology. Only rarely should cardiac catheterization be undertaken in children who have not previously undergone surgery for TOF with pulmonary stenosis. Catheterization carries a real risk of spasm of the infundibular outflow of the right ventricle induced by catecholamine and direct catheter, a complication that precipitates the need for emergency surgery, with the obvious increased risk relative to elective surgery [Jonas 2004]. Furthermore, the risk is increased in developing countries because the procedure is uncommon, proper instruments are lacking, and surgeons are inexperienced.

Three-dimensional echocardiography eliminates the need for geometric assumptions about LV geometry and therefore provides precise and accurate volume assessments [Gopal 1993]. Collection of a multitude of electrocardiographically gated sequential projections is cumbersome, however, and real-time 3-dimensional echocardiography is not yet a clinical reality. Assessment of local LV shape by using 2-dimensional echocardiography also has limitations, mainly because only a few projections are used to represent the whole LV [D'Cruz 1989]. A fault with the Nakata index is that it does not measure the subbranch segmental pulmonary arteries directly. The development status of the whole pulmonary artery cannot be evaluated by measuring the pulmonary artery diameter at only 1 point, usually just before first branch. In many TOF patients, postoperative outcomes were worse than predicted, even though their Nakata index was within the normal range. Because of the path of the left pulmonary artery and the limitation of visual angle, the proximal portion of the left pulmonary artery can usually be measured echocardiographically but the accuracy of the evaluation is affected. Magnetic resonance imaging provides anatomic and functional information that is superior to that provided by conventional cardiac imaging modalities such as echocardiography and heart ventriculography. Magnetic resonance imaging has its limitations, however, including long acquisition times that may necessitate the sedation of young children, and presents a problem in the examination of patients whose clinical condition is unstable [Boechat 2005].

Considering the insufficiency of the above diagnostic tools, additional information is necessary for a more comprehensive and accurate evaluation of pulmonary artery and LV function. Postural hypotension, a common phenomenon in people when they stand up from squatting position, includes a rush with dizziness, astasia, or even transient unconsciousness because of hypotensive cerebrovascular insufficiency. This phenomenon demonstrates that a significant and rapid decrease of systemic circulation resistance can occur simply from a position change. Squatting, a common symptom in TOF patients, improves arterial oxygen saturation by increasing systemic vascular resistance, thereby diminishing the right-to-left shunt and driving more blood flow to the lungs. In this process, right ventricular outflow obstruction is the most crucial factor for pulmonary blood flow. These phenomena are consistent with our hypothesis that the oxygen saturation difference between the different positions may reflect the development of right ventricular outflow.

In conclusion, it is our view that the SPO2 difference between the standing and squatting positions can reflect the potential function of the LV. Thus measurement of this difference may be an important accessory to LVEDVI in evaluating the prognosis of total correction of TOF with stenosis. Oxygen saturation and the extent of cyanosis reflect LV systolic function and pulmonary resistance. A limitation of this study was that patients with complicated TOF with pulmonary atresia and absent pulmonary valve were not included in this investigation. Further research and more detailed and comprehensive data about these complicated cases are needed.

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