A Case Based Approach to Calculating Shunts in a PAH Patient with Congenital Heart Disease

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Abstract: ***The most commonly used method to determine CO is the indirect Fick method (based on assumed rather than directly measured oxygen consumption VO2). In clinical practice, this method of CO determination yield inconsistent measurements in PAH patients and in congenital heart disease (CHD) population referred for RHC. Such possible inconsistencies in hemodynamic assessment may adversely impact clinical assessment and therapeutic decision-making. On the basis of a challenging case of a 42-year-old lady with a patent ductus arteriosus, we describe the approach for complex shunt volume, flows, flow ratio and resistance calculations based on the indirect Fick principle using hemodynamic data obtained in our catheterization laboratory.***

Keywords: Pulmonary arterial hypertension PAH, Right Heart Catheterization RHC, Fick method

1. Purpose

Pulmonary arterial hypertension (PAH) is a fatal and debilitating disease if not detected and treated early . A right heart catheterization (RHC) remains the gold standard in the diagnosis and evaluation of PAH. Determining the cardiac output (CO) is required to calculate the pulmonary vascular resistance (PVR). It is also an important prognostic hemodynamic parameter both in the initial evaluation and in follow-up.

The most commonly used method to determine CO is the **indirect Fick method** (based on assumed rather than directly measured oxygen consumption VO2). In clinical practice, this method of CO determination yield inconsistent measurements in PAH patients and in congenital heart disease (CHD) population referred for RHC.

Such possible inconsistencies in hemodynamic assessment may adversely impact clinical assessment and therapeutic decision-making.

2. Methods

On the basis of a challenging case of a 42-year-old lady with a patent ductus arteriosus, we describe the approach for complex shunt volume, flows, flow ratio and resistance calculations based on the **indirect Fick principle** using hemodynamic data obtained in our catheterization laboratory.

Clinical Vignette

A 42-year-old female patient, born to a first-degree consanguineous marriage, with no personal history of recurrent respiratory infections, was admitted to our institution for the assessment of a grade IV dyspnea and fatigue. She had no respiratory or cardiovascular symptoms until three years before her admission, with a progressive onset of dyspnea, associated with a paroxysmal nocturnal dyspnea, exertional angina and light headedness.

At physical examination, she was 68Kg. The heart rate was 90 bpm and blood pressure 110/50 mmHg. Saturation on room air was 85%, with chronic cyanosis, nail clubbing and conjunctival hyperemia. A fixed split second heart sound was heard on auscultation with no perceivable murmur. The patient also had a jugular venous distention, discrete peripheral edema and a positive Harzer's sign.

The gasometry showed hypoxemia (PaO2: 52mmHg) and normocarbia (PaCO2: 39mmHg) with oxygen saturation (SaO2) of 87%. **Hemoglobin** was 20 g/dL. **The ECG** showed regular sinus rhythm with right heart axis deviation, right ventricular hypertrophy, and negative anteroseptal and biphasic inferior T waves.

The Chest X-ray demonstrated cardiomegaly, elevated cardiac apex, enlarged right atrium and increased pulmonary vascular markings.

Echocardiography demonstrated right ventricular hypertrophy, a moderately dilated right atrium, enlarged pulmonary trunk, trivial tricuspid and pulmonary regurgitations, thus, pulmonary artery pressure evaluation was difficult. LV size and function were correct. A 10 mm patent ductus arteriosus was identified with a left-to-right shunt.

Right and left heart catheterization revealed a left-to-right shunt through the defect and severe isosystemic pulmonary hypertension (pulmonary artery pressure of: 131/73/101 mmHg). Hemodynamic data are presented in Table 2. **Aortic root injection** confirmed the presence of a large PDA.

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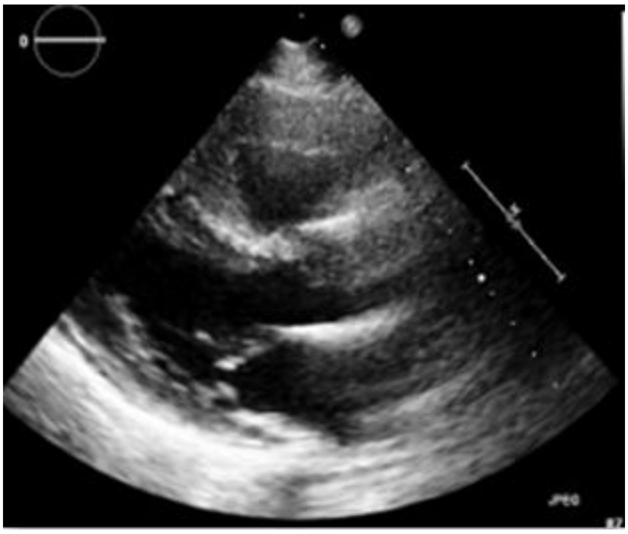
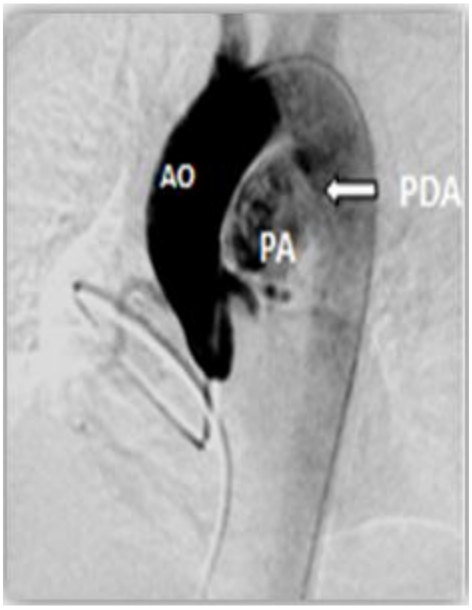


A: Parasternal short axis view shows biventricular hypertrophy predominantly right sided.

B: Hemodynamic data.



C: Lateral aortogram showing the PDA

3. Results

__**Fick Equation and Cardiac Output**__

- **Estimated VO2** at rest was calculated according to each of the 3 widely used empirical formulas listed in Table 1. In accordance with these equations, BSA was calculated according to **the formula by Du Bois**: BSA (m2) = 0.007184 × weight (kg)0.425 × height (cm)0.725.
- **Direct Measurement of** VO2 using the gold-standard technique of Douglas could not be performed due to the lack of material.

Dehmer	$\dot{V}O2$ (ml/min) = 125 (ml/min/m2) × BSA (m2)
LaFarge	$\dot{V}O2$ (ml/min) = 138.1 – (X × logeage) + (0.378 × HR) × BSA (Men: X = 11.49; Women: X = 17.04)
Bergstra	$\dot{V}O2$ (ml/min) = 157.3 × BSA + X – (10.5 × logeage) + 4.8 (Men: X = 10; Women = 0)

__**Cardiac Shunts**__ invalidate the concept of a single cardiac output, as there are different flows across the systemic and pulmonary circulations; inflow and outflow oxygen content varies depending on the site of sampling, the location of the shunt, and the direction of the shunt.

Using Fick's Principle, **three key Fick equations** are used to determine all important hemodynamic shunt parameters:

- **Systemic Flow (Qs)** is the total flow across the systemic circulation, **specifically between the femoral artery and the right atrium**.
- **Pulmonary Flow (Qp)** is total flow across the pulmonary circulation, **more precisely between the pulmonary artery and pulmonary veins**.
- **Effective Forward Flow (Qe)** is the theoretical **flow of unshunted blood through the entire circulatory system**. Sites of oxygen sampling are taken upstream to any shunting.

__**Shunt Flow**__

- **Recirculated pulmonary flow** is **the amount of left to right shunting**=fully saturated blood shunted to the right side of the heart **without passing through the systemic capillaries**.
- **Recirculated systemic flow** is **the amount of right to left shunting** =desaturated mixed venous blood that is shunted to the left side of the heart **without being oxygenated by the lungs**.

__**Pulmonary Arteriolar Resistance(PAR)**__ is the total resistance through the pulmonary arterial circuit and influences the degree and direction of shunting

In our patient

Cardiac output	VO2/ [(SaO2-SvO2)×1,36×Hb×10]	CO=2,66 l/min CI= 1,6 l/min/m2
Systemic flow	Qs=VO2/ [(PVO2-MVO2)×1,36×Hb×10]	2,66 l/min
Pulmonary flow	Qp=VO2/ [(PVO2-PAO2)×1,36×Hb×10]	13,3 l/min
Effective flow	Qe=VO2/ [(PVO2-MVO2)×1,36×Hb×10]	1,8 l/min
Recirculated pulmonary flow	Qp-Qe	11,5 l/min
Recirculated	Qs-Qe	0,86 l/min

systemic flow		
Pulmonary arteriolar resistance	[PAPm-PCWm/CO]×80/BSA	564n sec/cm5/m2, 564 15WU

Baseline summary: This patient has *bidirectional shunt* with a dominant left to right component of almost 11,5 l/min on room air (86% of Qp).

4. Conclusions

- Errors in hemodynamic assessment may adversely impact clinical assessment and therapeutic decision-making across a spectrum of serious cardiovascular conditions, especially cardiac shunts and PAH since shunt occlusion may lead to right ventricular failure and death for certain patients with severe PH.
- Recent papers suggest that **Thermodilution and Indirect Fick** are inaccurate methodologies to determine cardiac index in patients with Pulmonary Arterial Hypertension.

The indirect Fick method consequently underestimates CO in PAH patients, using an inaccurate estimation of VO2. In the other hand, predictive equations or assumed values do not accurately estimate oxygen consumption in the Congenital Heart Disease population, particularly in the setting of critical illness.

- The measurement of cardiac output using **thermodilution** typically requires the flow measurements to be done in circulations in a series with little to no shunt. The presence of a significant intracardiac shunt, therefore, renders this technique almost useless.
- While the **direct Fick method** is the most accurate method of determining flow, its applicability is limited. Most catheterization laboratories are not equipped or lack expertise to perform the direct Fick method.
- Non-invasive methods of measuring Qp and Qs have become more popular as technology has evolved. Doppler echocardiography and magnetic resonance imaging can be used to estimate Qp and Qs in patients with CHD.
- Those potential sources of inaccuracy could be avoided by calculation of the ratio of Qp to Qs will approximate the direction and magnitude of the shunting that is occurring, i.e., left to right (greater than one) or right to left (less than one), without quantifying flows (which would additionally require the measurement of systemic and pulmonary cardiac outputs).
- Studying variations under 100% O2or NO could also be helpful in decision-making.

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