

## “Cyanotic Blue Tongue” in Severe Rheumatic Tricuspid Regurgitation

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Central cyanosis is commonly associated with arterial desaturation, clubbing and a “blue tongue” [1]. On the other hand, the association of “blue tongue” with peripheral cyanosis is rare. We present a patient with this unique combination caused by long-standing severe rheumatic tricuspid regurgitation.

### Patient Description

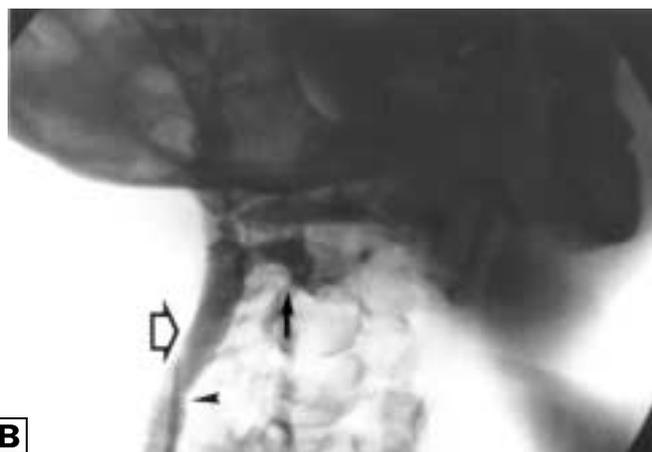
A 53 year old woman was hospitalized for progressive right-sided heart failure. Her past medical history revealed long-standing rheumatic heart disease, chronic atrial fibrillation, and mitral valve replacement in 1986. No extracardiac disorders were noted. Her recent medications included digitalis, diuretics and warfarin. The patient was cachectic, with mild proptosis, purple nose and cheeks, cyanotic lips and a “blue tongue” [Figure A]. No clubbing of the

fingers or toes was noted. Her resting blood pressure was 105/70 mmHg. The heart rate was irregular with a range of 75–80 beat per minute. She had a jugular venous pressure of 15 cm with a sharp Y descent, and precordial palpation indicated a right atrial and ventricular dilatation. Cardiac auscultation revealed irregular heart sounds, crispy closure and opening clicks of a prosthetic mitral valve, and a grade 3/6 left parasternal holosystolic murmur accentuated with inspiration. The liver was enlarged, pulsatile and tender. Both sacral and pedal edemas were detected. The electrocardiogram showed atrial fibrillation with an average rate of 75 beats/min, a frontal QRS axis of  $70^{\circ}$ , and clockwise rotation in chest leads with small Q and tall R waves in leads V1–V2.

Chest X-ray revealed both enlarged right atrium and right ventricle, a dilated

azygos vein and mild pulmonary venous congestion. Cardiac fluoroscopy showed preserved structure and function of the mitral valve prosthesis. On transthoracic echocardiography the patient had normal left ventricular size and function, moderate left atrial enlargement, and significant right atrial enlargement and right ventricular dilatation. Marked tricuspid annular dilatation (46 mm) was noted. Color flow mapping revealed severe tricuspid regurgitation. No right to left shunt at the atrial level was detected by intravenous right-sided agitated saline injection.

Catheterization of the right heart showed mean right atrial pressure of 18 mmHg with a V wave of 25 mmHg, right ventricular systolic pressure of 35 mmHg, and a mean wedge pressure of 14 mmHg. Pulmonary and aortic oxygen saturations were 65% and 95%, respec-



**[A]** “Blue tongue.” Note the engorged and dilated lingual veins (arrow). **[B]** Internal jugular venography (right anterior oblique view) shows a dilated and congested lingual vein (black arrow) and right internal jugular vein (open arrow). Arrowhead denotes catheter in right internal jugular vein.

tively. Cardiac output by the Fick method was 2.2 L/min. As shown in Figure B, venography of the great cervical veins depicted dilated and poorly drained internal jugular and lingual veins.

## Comment

Cyanosis is a bluish discoloration of the skin or mucous membranes resulting from an increased quantity of reduced hemoglobin or the presence of abnormal hemoglobin in red blood cells. This clinical sign may appear in various cardiac, pulmonary and hematological diseases [2]. In adulthood the central and peripheral types are the main clinical forms.

Cardiac right to left shunts or pulmonary diseases leading to systemic oxygen desaturation (i.e., <85%) are the main causes of central cyanosis. These forms usually occur in warm areas like the tongue and lips. In contrast, peripheral cyanosis is secondary to low output states associated with excessive oxygen extraction [3]. It generally occurs

in cool areas such as the nose, earlobes, cheeks and nail beds.

Long-standing severe rheumatic tricuspid regurgitation is hemodynamically characterized by a low cardiac output state, increased right atrial pressure and near-normal pulmonary pressures. This valvular abnormality is usually associated with peripheral cyanosis unless the foramen ovale is functionally patent or an intrapulmonary right to left shunt co-exists [4].

Our patient presented with typical signs of peripheral cyanosis, but had a “blue tongue” in the absence of systemic oxygen desaturation. This unusual finding can probably be explained as highly congested and dilated lingual veins. The dilated and congested lingual veins, as shown in Figure B, are secondary to increased right atrial pressure.

In light of the above, we suggest that severe tricuspid regurgitation leading to increased right atrial pressure should be added to the differential diagnosis of “blue tongue.” Moreover, finding a “blue tongue” in the presence of normal

systemic oxygen saturation should urge the clinician to look for severe tricuspid regurgitation.

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## References

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