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CASE REPORT

Blue in a Blink: A Telltale Tet Spell: An Uncorrected tetralogy of fallot in Hypercyonotic Crisis in the Adult: A Rare Encounter

Varshini D.R.[1] Balamurugan Nathan[2], Sandeep V. Joshi[3] Sreekara V.R.[4]

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ABSTRACT

Background: Tetralogy of Fallot (TOF) is recognized as the predominant form of cyanotic congenital heart disease beyond infancy. Grasping the dynamic factors that either exacerbate or ameliorate the shunt represents the cornerstone for managing critically ill patients with TOF in an emergency department (ED).

Case description: We herein document a 29-year-old uncorrected TOF patient who manifested symptoms of acute gastroenteritis & respiratory failure. A provisional diagnosis was then formulated connecting acute gastroenteritis to a provoked hypoxic spell with uncorrected TOF. Supportive care was provided that led to positive outcome.

Clinical relevance: This case study elucidates the clinical dilemmas in managing a critically ill, uncorrected TOF individual dealing with the complex interplay of physiology and biochemical changes. Attention to comprehensive management of a Tet spell is thereby underscored. Information suggests this situation represents a rare case of a few surviving uncorrected TOF patients documented in the Indian population.

KEYWORDS

• Cyanotic congenital heart disease • Fluid resuscitation • Uncorrected adult tetralogy of Fallot • Tet spell

AUTHOR'S AFFILIATION:

[1]Department of Emergency Medicine, Jawaharlal Institute of Postgraduate Medical Education and Research, Dhanvantri Nagar, Puducherry, India.

[2]Department of Emergency Medicine, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India.

[3]Department of Anesthesiology and Critical Care, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India.

[4]Department of Emergency Medicine, Jawaharlal Institute of Postgraduate Medical Education and Research, Puducherry, India.

CORRESPONDING AUTHOR:

Varshini D.R., Department of Emergency Medicine, Jawaharlal Institute of Postgraduate Medical Education and Research, Dhanvantri Nagar, Puducherry, India.

E-mail: Varshinidr66@gmail.com

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INTRODUCTION

The preponderance of deoxygenated blood within the capillary beds typically precipitates cyanosis, a noteworthy clinical sign. This particular clinical observation is primarily attributed to one of two pathological processes: either blood containing deoxygenated haemoglobin or the presence of abnormal haemoglobin forms incapable of delivering sufficient oxygen to end organs. Categorization of cyanosis recognizes two broad types: central and peripheral cyanosis. Central cyanosis stems from a reduction in pulmonary ventilation and oxygenation, impaired pulmonary perfusion, and the direct shunting of deoxygenated blood to the systemic circulation. Central cyanosis is best seen on perioral skin, oral mucosa, or conjunctivae and mostly secondary to profound arterial hypoxemia from hypoperfused state or cardiopulmonary disease including congenital heart disease. Referred to interchangeably as a hypercyanotic spell, the Tetspell constitutes a hallmark clinical manifestation of Fallot physiology involving hypoxic episodes, marked by paroxysms of rapid deep respirations and exacerbated cyanosis[1]as a result of total obstruction of the right ventricle outflow tract (RVOT).[2] Cyanosis becomes obvious during clinical assessment if the quantity of deoxygenated blood in circulation exceeds 5 g/dL. In infants, these episodes are frequently observed. The defining characteristics of Tetralogy of Fallot encompass (1) outflow tract obstruction, (2) overriding of the aorta, (3) ventricular septal defect (VSD), and (4) right ventricular hypertrophy. Patients diagnosed with Tetralogy of Fallot customarily exhibit polycythemia. In some instances, an individual with uncorrected TOF can remain asymptomatic until the onset of adulthood, if degree of RVOT obstruction is not severe. The occurrence of a tet spell in an adult signifies an exceptionally rare presentation in an ED setting a factor to consider.[18] We document the case of a 29-year-old man presenting with cyanotic spells, in whom a definitive diagnosis of TOF was diagnosed during early childhood.

CASE REPORT

This communication reports an instance of uncorrected TOF complicated by acute diarrhea and resultant shock, highlighting a consequential dilemma encountered during volume resuscitation efforts. Patient was not compliant to follow up clinical visits. He manifested hypercyanotic spells a few hours before presentation. He was brought to our ED with chief complaints of shortness of breath, lower abdominal pain, and loose stools since one day after ingesting food from a local resturant. There was no history of vomiting, bleeding per rectum, jaundice, altered sensorium. There was no history of food allergy or antibiotic abuse. There were no episodes of recurrent diarrhea in the past. At the time of presentation, he was concious & alert; recorded vital signs of respiratory rate of 40 breaths per minute, a heart rate of 140 beats per minute, and a recorded systolic blood pressure of 74 mm Hg. His room air oxygen saturation was not recordable via pulse oximetry. Arterial blood gas (ABG) results showed severe high anion gap metabolic acidosis. General physical examination identified an afebrile state, with cyanosis observed in the distal limbs and lips (*Figure 1A*) alongside pan-digitalclubbing (*Figure 1B*). Systemic examination revealed soft, non tender abdomen. Chest auscultation was unremarkable. Electrocardiogram disclosed sinus rhythm with a heart rate (HR) of 144 beats per minute, right axis deviation & incomplete right bundle branch block (RBBB) (*Figure 2A*). Bedside Ultrasound demonstrated a collapsed inferior vena cava and absence of B-lines in lung ultrasound. A provisional clinical diagnosis of Tetspell triggered by acute diarrhea in hypovolemic shock was formulated and treated accordingly. For initial stabilization, the patient was placed in propped up position, and oxygen therapy was supplemented via non rebreather mask at rate of 15L/minute. To manage the patient's clinical status, 10 ml/kg fluids were given in boluses; furthermore, vasopressor support and inotropic support were initiated to mitigate shunting and address septic shock. Phenylephrine was administered as bolus at 10 µg/kg dose, followed by an infusion at 3 µg/kg rate. Ketamine 30 mg was given intravenously. Sodium bicarbonate was infused at 1 meq/kg dose in parallel to address acidosis concurrently. Encouragingly, the patient

demonstrated improvement in vital signs, with progressive alleviation of symptoms.

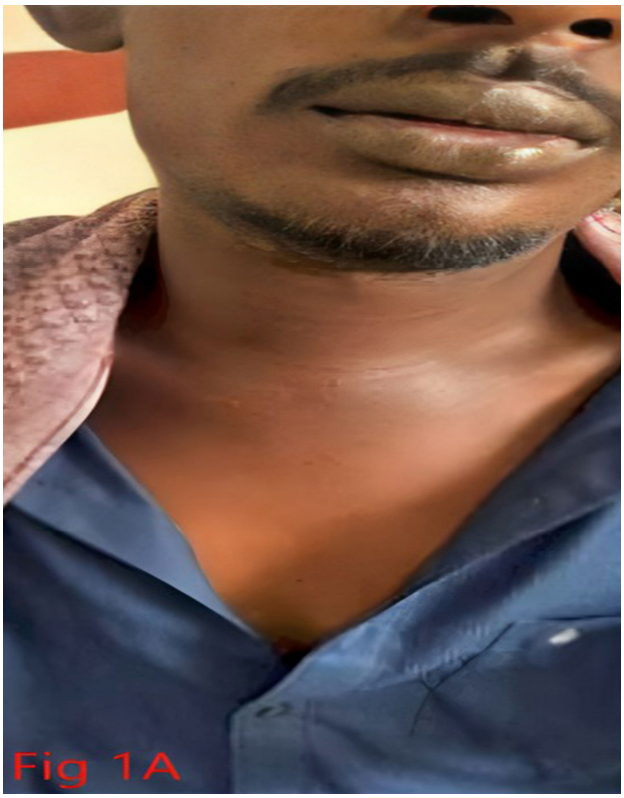


Figure 1A: Shows central cyanosis evident in the lips of an uncorrected TOF



Figure 1B: Illustrates Pandigital clubbing in an uncorrected TOF

Laboratory investigations revealed abnormalities in hemogram and renal function tests (*Table 1*). Leuococytosis, acute kidney injury, need for vasopressor therapy pointed towards septic shock rather than hypovolemic shock. Moreover, empirical antibiotics were adminstered after the procuring relevant cultures. This underscores the systemic implications documented.Chest radiograph pointed to pulmonary oligemia (*Figure 2B*). Corroboration for the TOF diagnosis was further established through a review of medical records. Two-dimensional echocardiography detailed normal left ventricular function, pulmonary atresia, overriding of the aorta, a D-shaped left ventricle, and a substantial VSD (*Figure 2C & 2D*). The patient's comprehensive evaluation pointed to acute gastroenteritis in septic shock, potentially a precipitant for this specific episode. In accordance with a targeted SpO2 of 70–75%, we elected to continue low-flow oxygen therapy of 4L/minute for an additional 24-hours.

Table 1: Various blood investigations obtained on arrival to ED

Hematological parameters	**Value**
Hemoglobin	23 g %
Leucocyte count	28,700 cells/cumm
Neutrophils/Lymphocytes	94% / 5%
Platelet count	89,000 cells/cumm
Urea	96 mg/dL
Creatinine	1.7g/dL
Sodium	122 meq/L
Potassium	5.4 meq/L
Arterial blood gas parameters	**Values**
pH	7.25
PCo2	28.4mmHg
PO2	39 mmHg
HCO3	12.2 mmol/L
Anion gap	28.6 mmol/L
SPO2	66%
Lactate	2.6mmol/L

He was subsequently transferred to a step-down unit on the fourth day of admission. Patient underwent one session of phlebotomy to manage hyperviscosity. Acute kidney injury resolved by the first week of admission. Following definitive cardiovascular evaluation, surgical repair was determined to be inadvisable for this patient; consequently, discharged on the eighth day of hospitalization.

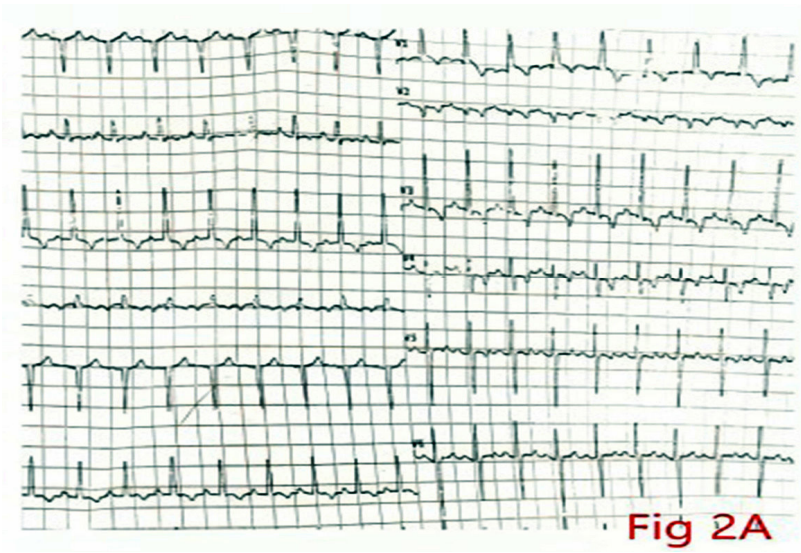


Figure 2A: Electrocardiogram showing sinus tachycardia of HR 144 bpm, right axis deviation, and RBBB. Also note the positive R wave in V1, suggesting pulmonary hypertension

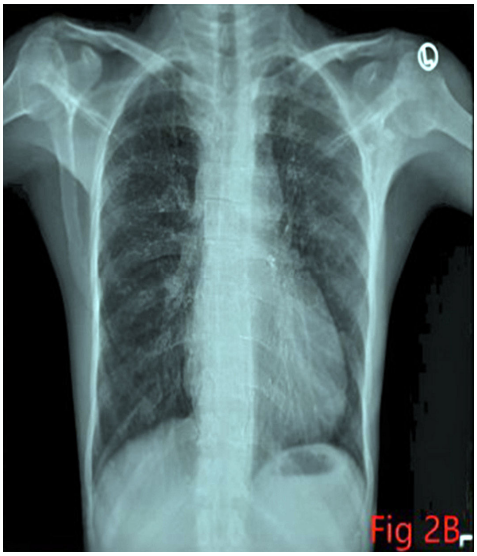


Figure 2B: Chest X-ray showing pulmonary oligemia in an uncorrected TOF

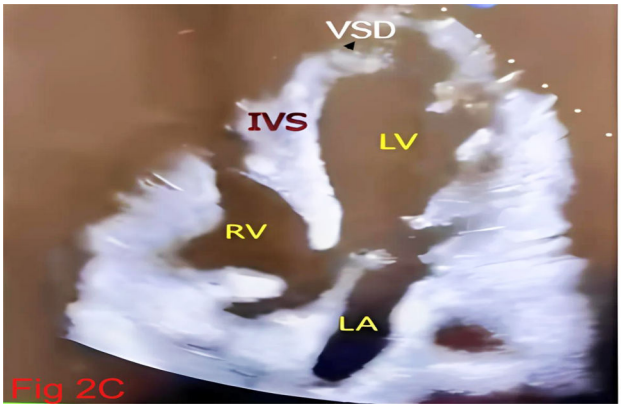


Figure 2C: Shows an echocardiography image of the index patient revealing a large VSD.

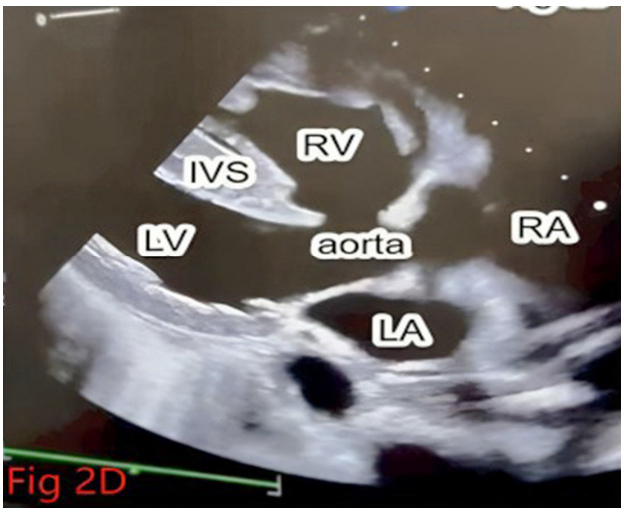


Figure 2D: Shows an echocardiography image of the index patient showing an overriding aorta arising from both ventricles. **RA** - Right atrium, **LA** - Left atrium, **RV** - Right Ventricle, **LV** - Left ventricle, Ivs-interventricular septum

DISCUSSION

This specific case serves as a poignant reminder regarding TOF as a key differential diagnosis for hypercyanotic crises, concurrently emphasizing its unique manifestations and intricate management.[3]

Specific adaptations are observed in adult TOF survivors, including hypoplastic pulmonary artery with the gradual origination of subpulmonary obstruction, extracardiac shunts.[4] It is understood that an unrepaired TOF, particularly when combined with mild to moderate RV outflow obstruction, can remain clinically subtle until adulthood. The case we encountered represented an emergent scenario, distinctly characterized by hypoxemia and a compromised ventilation-perfusion state. Diagnostic conclusions could nevertheless be facilitated by careful examination, which indeed revealed central cyanosis accompanied by clubbing in fingers. Clubbing in cyanotic congenital heart disease is attributed to right-to-left shunting, permitting megakaryocytes to bypass pulmonary fragmentation and reach the peripheries. Their cytoplasmic growth factor release initiates the proliferative changes responsible for clubbing.[5] Moreover, the minimal clinical improvement noted with supplemental oxygen further suggested a cardiac rather than primary respiratory etiology, raising strong suspicion of a **hypoxic spell** in the setting of chronic hypoxemia.

Mixing of blood occurs at VSD, with a right-

to left shunt enhancing deoxygenated blood concentration in the systemic circulation-making cyanosis manifest. The relative pressure gradient between the ventricles is the determining factor for the shunt. Any event instigating a reduction in systemic vascular resistance (SVR), such as defecation, hypovolemia, or tachycardia, will ordinarily produce a substantial right-to-left shunt across the VSD, thus initiating the onset of hypoxic spells.[6] It is classically understood that the large right-to-left shunt across the VSD circumvents the lungs, subsequently leading to a decrease in PaO2, an increase in PCO2, and a decline in the arterial pH. Following this, stimulation of the respiratory centers located in the brain, is triggered by these metabolic changes, eliciting hyperpnea. Such respirations amplify the negative intrathoracic pressure during inspiration, thereby augmenting the venous blood return to the right side of the heart.[7] This increased blood volume within the right ventricle is then shunted through the VSD, influenced by the combined effects of the existing right ventricular outflow obstruction and an acute decrease in SVR. This dynamic, one might ascertain, further perpetuates the hypoxic spell.The **strategic objectives of managing tet spells** are.[8]

- Preventing hypoxia
- Avoiding declines in SVR
- Abolishing hyperpnea
- Correcting acidosis
- Preventing hypothermia, tachycardia, and hypovolemia

Suppemental Oxygen: Supplemental oxygen should be provided either by mask or a nasal cannula. This measure improves oxygen saturation which concurrently reduces pulmonary vascular resistance (PVR).

Fluid Administration: Not only does hypovolemia precipitate cyanosis through a reduction in preload, which results in RVOT flow narrowing, but the R→L shunt also occurs, attributable to hypotension.Fluid boluses increase the blood volume in TOF patients with septic shock. Enhancing preload characteristically increases heart size; this, in turn, can potentially expand the diameter of the RVOT. Concomitant with the increase achieved in cardiac output and mixed venous saturation, fluid repletion effectively mitigates the risk of hypotension. Conversely, excessive fluid administration could precipitate cerebral edema, pulmonary edema, and hypoxia due to dilutional effects encountered in the setting of compensatory polycythemia.

Sedation: Any stress related tachycardia is effectively forestalled by sedation. Tachycardia instigates a proportional reduction in diastolic filling, thereby diminishing stroke volume while concurrently increasing myocardial oxygen demand. Thus, sedation helps reducing the work of breathing.

Morphine (0.1 mg-0.2 mg/kg) administered intramuscularly has been a traditional therapeutic option; however, it was avoided due to the possible untoward effect of systemic vasodilation through endogenous histamine release. Midazolam and fentanyl represent more contemporary alternatives.[9,10.] Regarding our index case, ketamine (1-2 mg/kg IV) was determined to be an excellent choice to substantively improve SVR, above and beyond its recognized sedative and analgesic properties[11,12] *Sodium bicarbonate*: In septic shock co-occurring with TOF, acidosis typically results in increased pulmonary vascular resistance and an augmented R→L shunt. In the setting of metabolic acidosis, sodium bicarbonate adminstration (1 meq/kg) may be implemented to break the vicious cycle of hypoxemia, acidosis, worsening hypotension

Vasopressors: For individuals whose condition does not improve with established measures, a vasopressor such as phenylephrine (5-20 µg/kg) may be administered slowly,[13] repeated every 10-15 minutes to escalate the SVR and thereby decrease the degree of shunt. Alternative options include methoxamine 0.1-0.2 mg/kg/dose IV. Prominent challenges are recognized in the management of uncorrected adult TOF patients manifesting with gastroenteritis and sepsis; these chiefly involve hypovolemia and a decline in SVR. Furthermore, determining volume status and guiding volume resuscitation in such a sepsis patient is frequently complicated, given that these patients typically demonstrate high hematocrit. Notably, an acceptable range for the patient's PaO2 was not reached, even with exposure to 100% FiO2, reflecting the nature of TOF as a cyanotic heart disease. A gradual reduction of FiO2 to a minimum of 21% was implemented over a 48-hours, predicated on

achieving his SpO2 target of 70–75%.

Our team diligently pursued shock resuscitation measures, grounding ongoing decisions in observed clinical improvement. A progressive tapering of vasopressor and inotropic supports was also subsequently undertaken.

As per current understanding, acute renal failure occurs infrequently.[14] However, it can manifest in hyper viscosity syndrome.[15] Cyanosis stimulates erythropoietin production, thereby augmenting red blood cells (RBCs) for enhanced oxygen-carrying capacity. Once hematocrit levels reach 65%, increased viscosity often ensues, leading to organ hypoperfusion and introduces coagulation abnormalities.[16] Chronic kidney disease has also been noted in patients with cyanotic congenital disease which is an effect attributed to hypoxia, hyperdynamic circulation, and polycythemia.[17] The development of thrombocytopenia can be attributed to either sepsis or attendant coagulation abnormalities.

We recommend fellow emergency physicians and critical care practitioners to monitor vigilantly for the potential emergence of acute kidney injury during tet spells. A robust interpretation of altered physiology in TOF, coupled with proficient management of septic shock, proves indispensable to emergency physicians for successful resuscitation. An overlooked diagnosis of this condition can result in compromised decision-making and inappropriate management; this frequently results in substantial morbidity stemming from complications tied to prolonged hypoxia.

LIST OF ABBREVIATIONS

ABG - Arterial Blood Gas analysis

ED - Emergency Department

RV - Right Ventricle

VSD - Ventricular Septal Defect

HR - Heart Rate

PVR - Pulmonary Vascular Resistsnace

RBBB - Right Bundle Branch Block

RBC - Red Blood Cells

RVOT - Right Ventricular Outflow Tract

SVR - Systemic vascular resistance

TOF - Tetralogy of Fallot

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