

## A unique case of uncorrected Fallot's tetralogy with nasal dermoid cyst and median cleft lip presenting during postpartum

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### ABSTRACT

While tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease among children, its first presentation in the third decade of life just after successful pregnancy outcome is extremely rare. In fact, survival of both child and mother having uncorrected TOF after noninstitutional delivery is unheard of. Herein, authors report a case of previously undiagnosed TOF associated with other midline congenital abnormalities, that is, nasal dermoid cyst and cleft palate, who presented for the first time with postpartum hemorrhage after an unsupervised home birth. To the best of our knowledge, this unique association has never been described before.

**Keywords:** Cleft palate, nasal dermoid cyst, postpartum, pregnancy, tetralogy of Fallot

### Introduction

While patients with tetralogy of Fallot (TOF) can nowadays survive up to adulthood due to the advent of corrective surgeries,<sup>[1]</sup> its first presentation during postpartum period and a successful pregnancy outcome is extremely rare.<sup>[2,3]</sup> Although TOF along with other congenital heart diseases (CHD) are frequently associated with multiple other congenital anomalies

including midfacial defects,<sup>[4]</sup> nasal dermoid cyst (NDC) in junction with CHD in an adult has never been described before. In this report, authors describe a unique combination of TOF in association with NDC and median cleft lip in a woman presented for the first time with postpartum hemorrhage (PPH) after an unsupervised home birth. A written informed consent was provided by the patient for publishing the medical records for academic purpose.

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### Case Report

A 30-year-old woman from low socioeconomic status presented with PPH following unsupervised non-institutional childbirth. Gynecologists arranged for blood transfusion and

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managed perineal tear that caused the PPH and the patient was transferred to department of medicine for unstable hemodynamic condition.

On general and cardiovascular system examination, she had features of cardiogenic shock. Other systemic examinations revealed no significant relevant abnormalities except median cleft lip, an NDC at the midline [Figure 1], and a subtle squint in left eye (all were present since birth). She also had history of exertional dyspnea since childhood and history of two spontaneous abortions. For aforementioned issues, she never sought medical attention.

Initial blood reports showed anemia (Hb—9.2 g/dL), neutrophilic leucocytosis (total count—36,700 with 87% neutrophils), raised liver enzymes (SGPT—221 IU/l, SGOT—389 IU/L), with renal profile suggestive of prerenal acute kidney injury (blood urea nitrogen—32.61 mg%, serum creatinine—1.27 mg%). Cardiac troponins were normal. Electrocardiography showed sinus tachycardia, right bundle branch block, and features suggestive of right ventricular hypertrophy (RVH). With the context of PPH, shock, sepsis, and an underlying suspected cyanotic congenital heart disease (CCHD) she was managed with noradrenaline infusion (@ 0.5–1 µg/kg/min), packed red cells transfusions, and broad spectrum antibiotics. Her hemodynamic status gradually improved and she was weaned off vasopressor support over the next 3 days.

2D-echocardiography showed situs solitus levocardia, large malaligned ventricular septal defect (VSD) with bidirectional shunt, RVH, overriding of aorta, subvalvular pulmonary stenosis with peak gradient of 32 mmHg with normal biventricular function suggestive of TOF [Figure 2a and 2b]. Computed tomography (CT) scans of thorax and abdomen were normal excepting the cardiac anomalies. CT scan of brain and paranasal sinuses revealed no intracranial communication of NDC [Figure 3].

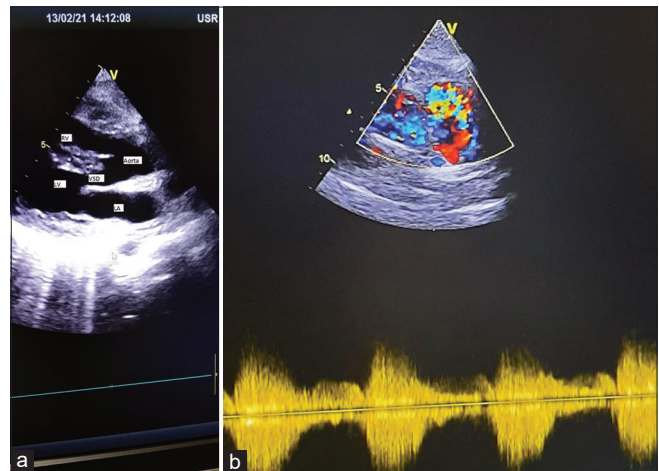
The newborn had low birth weight (1.5 kg) and poor cry, he was admitted for observation and was discharged healthy after negative screening for congenital anomalies. As the patient was hemodynamically stable at the seventh day post-admission, she was started with propranolol (20 mg/d) and torsemide (5 mg/d), followed by discharge and referral to higher center for possible corrective cardiac surgery. The couple was advised for contraception.

## Discussion

While TOF is the most common CCHD encountered in clinical practice, its first presentation in adulthood is rare.<sup>[5]</sup> Only 4% patients can cross third decade of life with uncorrected TOF.<sup>[6,7]</sup> There are various hypotheses to explain survival upto adulthood with uncorrected TOF. Initially mild but eventually progressive pulmonary stenosis (counterbalancing the left



**Figure 1:** Nasal dermoid cyst and median cleft lip



**Figure 2:** (a) 2-D echocardiographic parasternal long-axis view showing large malaligned VSD with overriding of aorta. (b) 2-D echocardiographic parasternal short-axis view at basal level showing subvalvular pulmonary stenosis with peak gradient 32 mmHg and confluent good size branch pulmonary arteries



**Figure 3:** CT scan PNS shows subcutaneous predominantly fat density (of - 95 HU) lesion overlying the nasal bone without any intraosseous or intracranial communication

ventricular load and protective for pulmonary vasculature), formation of systemic to pulmonary collaterals (both cardiac and extra-cardiac), concomitant systemic hypertension, and polycythemia have been proposed as probable defensive mechanisms.<sup>[2,8-10]</sup> In the present case, only the first factor has probably played its role.

Pregnancy in TOF carries heightened risk of both maternal morbidity and mortality along with poor perinatal outcome.<sup>[11-13]</sup> The risk is even greater among uncorrected TOF cases as pregnancy is associated with decreased peripheral vascular resistance, increased pulmonary vascular resistance resulting in increased right to left shunt with more hypoxia.<sup>[14]</sup> Evidence of successful pregnancy outcome in uncorrected TOF without any antenatal checkup is limited in literature.<sup>[3,14]</sup> There are reports describing favorable fetomaternal outcome among uncorrected TOF patients with intensive obstetric, anesthetic, and cardiac care.<sup>[3,15,16]</sup> Our patient was unique as she received no medical attention till she presented with PPH, heralding cardiogenic shock. This also highlights dismal healthcare access and awareness among poor tribal people of the region.

Multiple craniofacial defects have previously been described in TOF including hypertelorism, micrognathia, oculoauriculovertebral dysplasia, hemifacial microsomia, and velopharyngeal insufficiency.<sup>[4,17-19]</sup> Cleft lip and palate have been described as the potential clinical clue for underlying CHD.<sup>[20]</sup> Had the index patient come in contact with healthcare before, she should have been screened for CHD.

Midline nasal mass present since birth in this patient could be encephalopcele, dermoid and epidermoid cysts, glioma, hemangioma, lymphangioma, and dacryocystocele.<sup>[21]</sup> Clinico-radiologically it was diagnosed as dermoid cyst due to its location, presence of tuft of hairs, and radiological exclusion of other entities.<sup>[21]</sup> NDC comprises 1% of all dermoid cysts, mostly come across before the age of 3 years.<sup>[22]</sup> Adult NDC discovered incidentally among adult as in the present case is extremely rare. It is important to note that NDC can present as isolated superficial cystic lesion (type-I/nasal surface type) or may be associated with fistula or sinus encroaching nasal bones (type-II/nasal intraosseous type) extending into deeper intracranial structures (type-III/intracranial epidural type and type-IV/intracranial dural type).<sup>[22,23]</sup> Absence of intraosseous/intracranial extension prevented complications. Besides, extreme rarity among adults, NDC has never been described in association with TOF.<sup>[24]</sup> Thus, this report certainly expands the spectrum of facial defects among CHD described in literature. Moreover, the occurrence of NDC with median cleft lip is also very rare. While NDC occurs due to embryological defect in frontonasal region, the median cleft lip probably occurs as a secondary event due to non-fusion of frontonasal process and maxillary process owing to presence of NDC in between.<sup>[25]</sup> She refused when excision of the cyst followed by rhinoplasty was offered as treatment.

## Conclusion

Index presentation of a CCHD (TOF in cardiogenic shock) in postpartum period following successful pregnancy outcome and its association with another rare midfacial defect (NDC and median cleft lip) makes this case unique. Cardiac and brain magnetic resonance imaging, cardiac catheterization, and genetics studies could not be done due to lack of infrastructural and

logistic backup. In cases of any congenital midline anomalies, even in the apparent absence of previous historical clues over the decades, CHDs should be carefully sought for.

## Declaration of patient consent

Informed consent taken from the patient and attendant for using the medical records for academic purpose.

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## Conflicts of interest

There are no conflicts of interest.

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