INNOVARE JOURNAL OF HEALTH SCIENCES

Vol 8, Issue 5, 2020

ISSN - 2347-5536

Case Report

UNOPERATED TETRALOGY OF FALLOT: A CLINICOPATHOLOGIC STUDY OF A 12 MONTHS CHILD AT GOVERNMENT GENERAL HOSPITAL, KADAPA

SHAIK KAREEMULLA, HARINI N*, PRASANNA REDDY V, SAI PRASANNA K

Department of Pharmacy Practice, P. Rami Reddy Memorial College of Pharmacy, Kadapa, Andhra Pradesh, India.
Email: harinihasini760@gmail.com

Received: 24 February 2020, Revised and Accepted: 27 March 2020

ABSTRACT

Tetralogy of fallot is a congenital cardiac malformation that consists of an interventricular miscommunication. This combination of lesions occurs in three of every 10,000 live births and accounts for 7–10% of all congenital cardiac malformations. It is a condition caused by a combination of defects in all the four heart chambers that are present at birth. These defects affect the structure of heart, cause oxygen poor blood to flow out of the heart and to the rest of the body. Infants and children with TOF usually have blue tinged skin. The prevalence rate of TOF is 3.5% affecting males and females equally. Pneumonia is an infection of lungs that inflames air sacs in one or both lungs. The air sacs may fill with fluid or pus causing cough with phlegm, fever, and chills. It is most common in infants and young children and people with weakened immune system. A 12 months' female child with this congenital heart defect and pneumonia was discussed in this report. However, most of the children with TOF need surgery, but timings may vary depending on the condition and severity. This is important to note that surgery for TOF is palliative but not curative. This report concludes that there is a need to increase awareness of TOF so as to encourage early diagnosis and therefore promotes better outcomes.

Keywords: Tetralogy of fallot, Emergency care, Pulmonary cyanosis, Heart murmurs.

INTRODUCTION

Tetralogy of fallot (TOF) is a congenital heart defect that occurs at birth. TOF was first described by Niels Stenson, Danish anatomist in 1672. Sandifort Edward describes the symptoms and anatomical findings of TOF in a child in 1777 [1]. Significant congenital heart disease may be diagnosed virtually at any age but some conditions are discovered in rarely identified in infants [2]. It is one of the most common causes of cyanotic congenital heart diseases. It accounts for 7–10% of all congenital cardiac lesions and affects males and females equally [3]. The prevalence of TOF is 3.5% and the child born with TOF in these days can expect to survive to adulthood because of advances in the surgical treatment [4]. Congenital heart disease is a gross structural abnormality characterized by ventricular septal defect (VSD), over-riding aorta, pulmonary stenosis, and hypertrophy of right ventricle.

TOF has many causes. The major cause is chromosomal abnormalities accounts for 25% of cases of child with TOF. It includes triosomes 13, 18, 21, and 22q 11.2. Among these, most common abnormalities are triosomes 21 and 22q 11.2. Moreover, other causes are family history, Digeorge syndrome [5]. The common risk factors associated with TOF includes viral infections, measles during pregnancy, and maternal intake of retinoic acid in first trimester, untreated phenylketonuria, and poorly controlled diabetes mellitus in pregnancy [1,5].

However, onset of symptoms and severity depends on degree of right ventricular outflow tract obstruction. In mild cases, the patient is asymptomatic and acyanosed. The only finding present in such cases is systolic murmur. In case of severe obstruction, marked cyanosis may be present which requires medical intervention. Signs and symptoms become more apparent as the infant grows older. Along with the cyanosis, respiratory distress and hyper-cyanotic spells (TET spells) may also present [1,5]. These TET spells are life threatening and may be fatal which requires intervention. It occurs as a result of spasm of right outflow tract. Feeding, crying, and exertion are the precipitating factors of TET spells.

The pathology of TOF includes conal septum is displaced anteriorly. This displaced conal septum protrudes into the pulmonary outflow tract, which may result in obstruction and blockage of downstream structures including the pulmonary valve, main pulmonary artery, and branch pulmonary arteries. Systolic pressure in the left and right ventricles and in the aorta is same because VSD is large. The extent of condition depends on the degree of right ventricular outflow obstruction. This obstruction may result in a net left to right shunt through the VSD, and a severe obstruction leads to a right to left shunt resulting in low systemic arterial saturation. TOF is majorly confirmed by echocardiography. Clinical diagnosis requires history, and clinical examination. Chest X-ray is done to identify boot shaped heart. Right ventricular hypertrophy can be identified by electrocardiography. Cardiac catheterization can be done in case of suspicion of any coronary anomaly. TOF condition is manageable by medication and surgery. The mode of management depends on type and degree of right outflow obstruction. In case of critical pulmonary stenosis, prostaglandin infusion (0.05–0.1 mcg/kg/min intravenous [IV]) is given to increase pulmonary outflow of blood. Surgical intervention is necessary in case of severe right ventricular outflow. It includes intra-cardiac repair and shunt/bypass surgery [6]. For treating TET spells, calm the child and provide supplemental oxygen and infuse IV fluids for volume expansion. If the spell persists, standard therapy should be followed. It includes beta-blockers (propranolol or esmolol), morphine, phenylephrine, intranasal midazolam, and intranasal fentanyl.

PNEUMONIA

Pneumonia is a respiratory condition in which there is an infection of the lungs that inflames the air sacs. Globally, pneumonia sustains as the leading cause of death of children under 5 years. According to the WHO report, pneumonia accounts for 120 million cases every year, among which 14 million cases were severe [7]. Pneumonia is caused by variety of organisms. The most common causes are bacterial (*Streptococcus pneumonia*, and mycoplasma), virus (flu virus, adenovirus, parainfluenza virus, and respiratory syntical virus), and fungus (*pneumocystis jiroveci*). The group of people who are at risk of long-term morbidity and mortality is infants (low birth weight or premature), those people who are immunocompromised, and those who have other conditions such as cystic fibrosis and congenital heart disease. Child can present signs and symptoms at different stages of illness. Clinical presentation

of pneumonia includes fever, cough, tachypnea, wheezing, abdominal pain, and lethargy. Other signs of pneumonia are chest indrawing, nasal flaring, cyanosis, breathing difficulty, feeding less than half of normal intake, and signs of dehydration. A temperature of >38.5°C is a characteristic feature of bacterial pneumonia [8].

Pathology of pneumonia starts with invasion of lower respiratory tract by inhalation, aspiration of pathogens. To counteract the pathogens, the barrier to infection anatomical structures includes nasal hairs, cilia, epiglottis and cell mediated, and humoral immunity activated. As a result of these, nasopharyngeal colonization occurs which leads to inflammation or injury or death of surrounding epithelium and alveoli. The migration of inflammatory cells to the site of infection causing an exudative process that impairs oxygenation [9]. A number of different laboratory tests ate used currently in combination with the clinical assessment to diagnose pneumonia. White blood cells (WBC count), C – reactive protein, and erythrocyte sedimentation rate are generally used as a markers of infection, but they do not help in distinguishing between bacterial, viral, or mixed pneumonia. Blood cultures are taken if suspected to have bacterial pneumonia. Polymerase chain reaction enhances the identification of specific organism, but it is not widely available, expensive and routine use is not recommended. Viral immune-fluorescence and viral antigen detection can be done by nasopharyngeal aspirate which is useful in identifying virus [8,10].

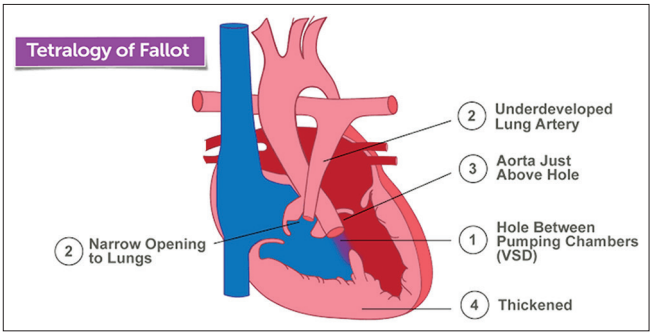

Fig. 1: Tetralogy of fallot with pulmonary atresia

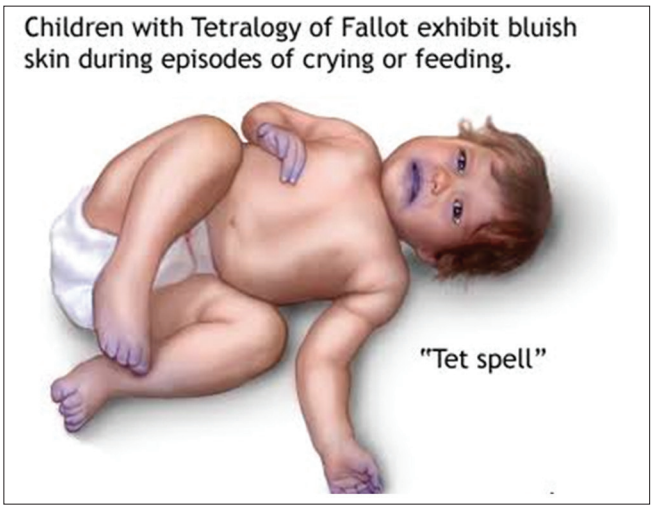

Fig. 2: Congenital cyanotic heart disease

Chest-X ray is still considered as a golden standard for diagnosing pneumonia. In general, it is not possible to identify the exact cause of pneumonia in the clinical practice. This is because of coinfections of bacteria and virus which cannot be distinguished from infection due to a single pathogen. Hence, antibiotic therapy is prescribed for all the children. The main goal of treatment is to reduce the risk of morbidity and mortality of pneumonia as cheaply as possible. In severe pneumonia, the recommended first-line drugs are benzyl penicillin, amoxicillin, and chloramphenicol. In case of very severe pneumonia, ampicillin, or amoxicillin plus gentamicin are recommended [11]. Amoxicillin is the first choice for children <5 years old through oral route, alternative are coamoxiclav, cefaclor, erythromycin, clarithromycin, and azithromycin. At present, resistance of *S. pneumonia* to penicillin is increased. Hence, macrolides are prescribed along with it. The British Thoracic Society guidelines suggested that antibiotics administered orally are safe and effective and IV antibiotics are preferred for children with severe signs and symptoms. In general, oral antibiotics are prescribed for 5–7 days but treatment duration is increased to 10 days based on severity of disease and antibiotic. The major complications of pneumonia are treatment failure, pleural effusion, emphysema, lung abscesses, septicemia, etc. [8]. The hospitalization and mortality of pneumonia can be reduced by conjugate vaccines against *Haemophilus influenza* type b and *S. pneumonia* [12].

A CLINICO-PATHOLOGIC STUDY

A 12 months' female child patient weighing 4.5 kg, resident of kadapa was admitted in PICU ward presented with chief complaints of fever, cough, cold, and breathlessness since 4 days. The patient's mother noticed bluish discoloration of fingers, toes, lips during irritability, and

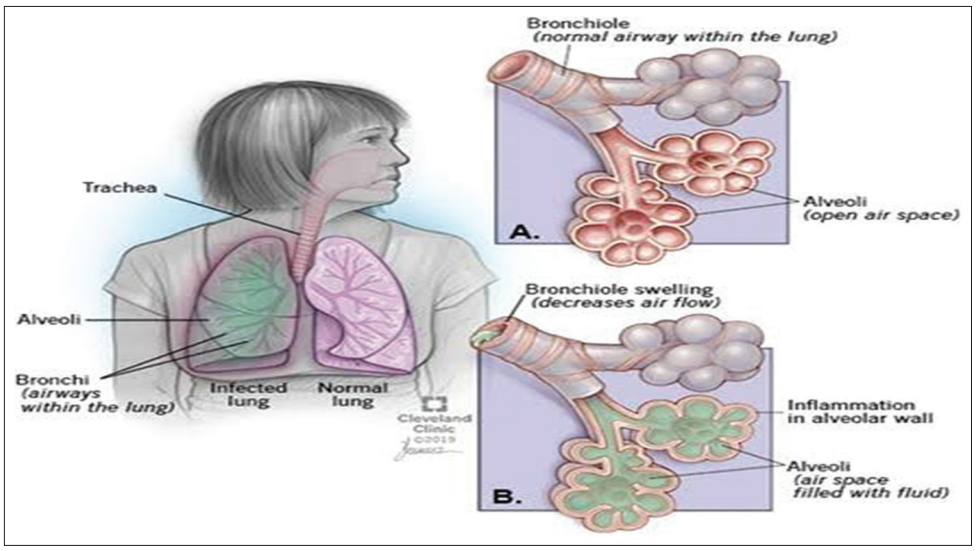

Fig. 3: Bronchi swelling and alveoli filling with mucus

Table 1: The patient was treated with following medications

Drug name	Dose	ROA	Frequency	Duration (days)
Inj. Piptaz	500 mg	IV	TID	1–17
Inj. Amikacin	40 mg	IV	OD	1–17
Syp. Ambroxol	1 ml	p/o	TID	2–15
Syp. Paracetamol	2.5 ml	p/o	TID	1–3
Neb Budecort	0.3 mcg	Nasal	12^{th} hrly	1–17
O_2 Inhalation	5 L/min	Nasal	SOS	1–20
Furoped drops	0.5 ml	p/o	BD	1–20
Isolyte– P	150 ml	IV	TID	1–14

excessive crying. Medication history of the patient includes, TOF was observed immediate after 10 days of birth. Medication history includes Tab. Propranolol 4 mg TID at the age of 9 months. In addition, patient was prescribed with syrup Digoxin 12.5 μg (0.25 ml) BD since 1 month. Patient was immunized up to the age as per schedule. She was born of a full-term cesarean delivery in the hospital without any peripartum complications but with low-birth weight of 1.77 kg and not cried immediately after birth. Personnel history includes weight – 4.5 kg, abnormal sleep, bowel, and bladder habits that were normal, appetite was normal. There was no history of similar illness in the family.

On examination, the patient was found to be conscious, coherent, and cooperative. She was comfortable at rest. The pulse rate was 118/min, respiratory rate was 62 cpm, temperature was 100°F. The digits of all four limbs and lips are cyanosed and a saturation of 58%. On cardiovascular examination, S1 and S2 were normal. Pan-systolic murmur was heard, loudest. On respiratory examination bilateral coarse crepts also present. Chest X-ray report shows consolidation in both lungs at basal fields and liver was not palpable. 2D Echo reveals double outlet right ventricle, TOF, straddling of tricuspid valve, and mild-to-moderate hypo-plastic left ventricle. During the treatment course, the patient oxygen saturation was reduced day-by-day from 70% to 35%.

Gradually, the patient condition became worsening and dobutamine infusion of 1.35cc was given on days 18 and 19. The patient passed away on day 20 without undergoing surgical intervention.

DISCUSSION

This is a case of an infant with TOF and bronchopneumonia. In general, the diagnosis and clinical management of TOF requires multidisciplinary team that includes a pediatrician, pediatric cardiologist, cardiac surgeon, radiologist, and pharmacist. However, most of the children with TOF need surgery, but timings may vary depending on the condition and severity [13]. This patient undergone symptomatic therapy for TOF and hyper-cyanotic spells since 10 months and patient passed away at the age of 12 months. She does not undergo any intervention. There is 75% mortality rate for child with TOF without any intervention and 70% survival rate for child with TOF who undergone corrective surgery either in childhood or adulthood [13].

In US, infants born with TOF undergoes primary repair within 1^{st} year of life and outcomes were good but after 20 years majority people need pulmonary valve replacement. Nowadays, percutaneous method of implanting pulmonary valve is available, in which child is free from symptoms but the long-term results were unknown. This is important to note that surgery for TOF is palliative but not curative [1]. This case demonstrates the challenges faced by the child with cyanotic congenital heart disease without any intervention. Hence, this report concludes that there is a need to increase awareness of TOF so as to encourage early diagnosis and therefore promotes better outcomes. However, pharmacist also plays a role in management of TOF by reducing medication errors, drug-drug interaction, improvement of usage of guideline driven therapy, and promotes medication adherence [14].

ACKNOWLEDGMENT

We wish to express our gratitude to a number of people who made important contributions to the accomplishment of this case report. It is pleasure to convey our gratitude and thank health care staff of Rajiv Gandhi Institute of Medical Sciences, Government General Hospital, Kadapa, Andhra Pradesh, for valuable help with the literature collection, supervision, advice, and guidance throughout the work.

COMPETING INTERESTS

We declare that we have no competing interests.

AVAILABILITY OF DATA AND MATERIALS

This is a case report.

ETHICAL APPROVAL

Not required.

REFERENCES

1. Diaz-Frias J, Guillaume M. Tetralogy of Fallot. Treasure Island, FL: Stat Pearls; 2019.
2. Silberbach M, Hannon D. Presentation of congenital heart disease in the neonate and young infant. Pediatr Rev 2007;28:123-31.
3. Motiram R, Shankarrao V. Prevalence, profile, and pattern of congenital heart disease in central India: A prospective, observational study. Niger J Cardiol 2018;15:45-9.
4. Hu BY, Shi K, Deng YP, Diao KY, Xu HY, Li R. Assessment of tetralogy of Fallot-associated congenital extracardiac vascular anomalies in pediatric patients using low-dose dual-source computed tomography. BMC Cardiovasc Disord 2017;17:285.
5. Animasahun BA, Madise-Wobo AD, Falase BA, Omokhodion SI. The burden of fallot's tetralogy among Nigerian children. Cardiovasc Diagn Ther 2016;6:453-8.
6. Gillani SW, Baig MR, Sari Y, Nuraihan S, Razali B, Siddiqui AA. Case report clinical evaluation of treatment for congenital heart defects (ASD and VSD) with bronchopneumonia. Int J Pharm Life Sci 2014;5:28-53.
7. Jahan Y, Rahman A. A case report on management of severe childhood pneumonia in low resource settings. Respir Med Case Rep 2018;25:192-5.
8. Lakhanpaul M, Atkinson M, Stephenson T. Community acquired pneumonia in children: A clinical update. Arch Dis Child Educ Pract Ed 2004;89:29-34.
9. Ebeledike C, Ahmad T. Paediatric Pneumonia. Treasure Island, FL: Stat Pearls; 2019.
10. Rodrigues CM, Groves H. Community-acquired pneumonia in children: The challenges of microbiological diagnosis. J Clin Microbiol 2018;56:e01318-17.
11. Principi N, Esposito S. Management of severe community-acquired pneumonia of children in developing and developed countries. Thorax 2011;6:815-22.
12. Roux DM, Zar HJ. Community-acquired pneumonia in children-a changing spectrum of disease. Pediatr Radiol 2017;47:1392-8.
13. Aluri K. Tetralogy of fallot without corrective surgery in a 33 year-old ICU patient. Pediatrics 2017;140:35-9.
14. Cheng JW. Current perspectives on the role of the pharmacist in heart failure management. Integr Pharm Res Pract 2017;7:1-11.