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Holistic Care in a Baby with Prolonged Hospitalization in Pentalogy of Fallot: A Case Report

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ABSTRACT

Pentalogy of Fallot is a rare congenital heart defect comprising of classic four features of Tetralogy of Fallot (TOF): ventricular septal defect, pulmonary stenosis, overriding aorta and right ventricular hypertrophy with addition of an atrial septal defect or patent ductus arteriosus. An additional ASD is seen in 3 to 5% of children with TOF. Treatment comprises of surgical correction of the defect involving patch closure of the ventricular septal depict, resection of the infundibular stenosis, and pericardial patch to decrease the RVOT obstruction. The following case represents a baby with pentalogy of Fallot who underwent surgical correction of the defect and received holistic treatment (surgical, nursing, dietary, physiotherapy) in a prolonged hospitalization (1 year) following the surgery to alleviate the disease condition of the baby.

Keywords: *pentalogy of Fallot, holistic care, prolonged hospitalization, tetralogy of Fallot*

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BACKGROUND OF THE STUDY

A 22 days old neonate presented in emergency department of AIIMS on nasal prongs O_2 at 5 l/min and cyanosis (peripheral cyanosis), fast breathing (65 breaths/min) present, HR: 130 beats/min, BP 95/36 mmHg, active baby, was full term normal vaginal delivery on 22/2/2019, cried at birth, APGAR of 9 at 5 min, passed meconium, completely normal at birth, no family history of congenital cardiac disease, with bluish discoloration of extremities noted after 9 days after birth, no suck-rest-suck cycle, evaluated at UP and further referred to higher centre. Neonate was primarily admitted in emergency department, AIIMS.

Pre-Operative Notes

Baby was stabilized with PGE1 infusion (PGE1 0.01 mcg/kg/min) and was diagnosed with Pentalogy of Fallot, following an ECHO; and CXR revealed oligemic lung fields**.** Decreasing levels of saturation (65–70%) necessitated intubation. Abdominal distension and melena (4 times) was noted on 2nd day after admission, nasogastric tube was inserted which drained 75 ml of greenish aspirate. Abdominal USG ruled out Ischemic ileus, septic ileitis and malrotation - showed sluggish movement,

minimal free fluid in abdomen. Blood findings reported thrombocytopenia (26,000) and INR of 1.7, indicating NEC Stage 1b. Baby was kept NPO, started on IV fluids and aminoven (1.5 ml/h), an antibiotic cover of amikacin, ciprofloxacin and cefotaxime and Vit. K weekly. NEC resolved on day-5 of admission, abdominal sounds resumed and feeds were started @10 ml/3 hourly, tolerated well. The baby underwent duodenal clipping on 27/3/2019 due to frequent melena episodes.

Child's condition stabilized later and was posted for surgical correction of defect, underwent *ICR+RVOT resection + Direct suture closure of ASD+PDA clipping on 28/3/2019.*

Post-Operative Notes

Baby was kept on PRVC with controlled ventilation on 1st POD, on 2nd POD, in view of decreased urinary output, baby was put on peritoneal dialysis (PD fluid+50% dextrose), continued till POD 8. Difficulty in weaning off the baby from ventilator was noted with consequent need of positive pressure. On POD 20, baby was extubated but reintubated on same day following drop in saturation (55%) and drop in heart rate (70), subglottic stenosis was noticed at time of reintubation. On further investigations, bronchomalacia was also noticed during bronchoscopy. Frequent nebulization was advised every 4th hourly and *suctioning* (*only when necessary* in view of thick mucoid secretions), due to excessive secretions and frequent bronchospasms. Baby was advised for limb physiotherapy every 6 hourly. The child was given various trials for extubation thereafter, kept on SIMV-CPAP modes of ventilation. On decreasing SIMV support subsequently, child presented increased distress. During CPAP mode, ventilatory demand was decreased to T Piece and CPAP (4:½ h) during a span of 2 months after surgery. Child had difficulty on frequent intervals requiring increased PEEP and PS support. On CXR, the right diaphragm was elevated; suggestive of *Right diaphragmatic palsy*, increased respiratory distress off ventilator, with left hyperinflation and increased crepts on auscultation.

Post-Operative Day: 124

In view of inability to wean off from ventilator and recurrent respiratory distress:

Bronchoscopy

Granulation tissue over trachea, B/L bronchomalacia (advised for ventilation with CPAP, high PEEP and controlled trials of extubation). Tried extubation but baby had bradycardia, upward rolling of eyeballs, no saturation drop, thus put back to SIMV support.

Diaphragmatic Plication-I (30/7/19)

Child was again subsequently weaned off to CPAP, and was put on mother's lap under controlled guidance for a day, but on second day, baby developed bradycardia (56) episode which was resolved by cardiac massage for 4–5 sec.

Direct Laryngoscopy (14/8/19)

Child had bradycardia episode with need of CPR while shifting to mother's lap. To find out causes of inability to wean off, procedure was done and granulation tissue was removed which revealed right lower collapse and bilateral bronchomalacia.

Diaphragmatic Plication-II (25/9/19)

Baby underwent procedure for second time and presented with consequent three episodes of bronchospasm in a 24 h of span and episode of desaturation followed by bradycardia which resolved on CPR for 2 min. Next day, the baby went drowsy and developed gasps. Baby presented with vomitus in mouth, chest compression and

bag and mask ventilation started and emergency drugs were given, baby revived (was started on phenobarbitone and levera).

Shifted to Ward from NICU on 24/11/19 on Subsequent SIMV: CPAP mode FiO_2 60% and Increased Pressure Support

Child was put on NG Tube feeding in view of GERD and was kept propped up.

a. Strict observation;
b. Immediate initiation of Bag-Mask Ventilation (in case of desaturation);
c. Gentle tracheal suction (5–6 cm);
d. Nebulisation with Adrenaline and Duolin x Q3H; and
e. CPT, 2nd hourly position.

Child was also kept on drip feeds (November to December) in addition to normal feeds to increase weight and put on continuous cardiac monitoring, with hourly monitoring of vital parameters, involvement of parents in care, physiotherapy and suctioning as per need. During stay of child a proper feed management of child was done throughout as per age and condition of baby. Weight of baby increased from 2.9 to 3.8 kg to a maximum of 5.9 kg. Management of baby had various difficulties due to multiorgan involvement. Though child was not totally stable but discharged and shifted to centre near their native place on basis of difficulty of parents who had been staying in Delhi for more than a year.

INVESTIGATIONS

Pre-Operative ECHO (13/3/20)

Hypertrophied RV, pulmonary atresia, with aneurismal IAS, small RV, confluent B/L pulmonary arteries (3 mm each), hypertrophic tripartite RV, hypoplastic PVs (4.5–4.8 mm), small PDA.

CT for Post-Operative CHD (1/6/19)

Normal situs, MPA: 4.4 mm (proximal to bifurcation); RPA: 3.3 m; LPA; 4.5 mm; DTA: 4.8 mm; PDA absent no significant aortopulmonary collaterals, from DTA seen, collapse of right lower lobe, left lung normal.

CT for Post-Operative CHD (29/8/19)

Atria: residual ASD+, MPA: 6.6 mm (proximal to bifurcation; RPA: 4 mm; LPA: 3.7 mm, DTA: 6.3 mm, few significant aortopulmonary collaterals, confluent PAs. Lung parenchyma and mediastinum collapse—consolidation of basal segments right lower lobe noted. Segmental atelectasis seen in left lower lobe.

NCCT Chest (14/8/19)

Compression of airway with segmental collapse consolidation of bilateral lungs.

NCCT Chest (11/11/19)

Consolidation involving the superior segment LLL with volume loss of left lung.

Blood Investigations

CBC revealed a haemoglobin concentration ranging from mostly around 10 gm/dl (with frequent transfusions to keep Hb>10), platelet concentration of minimum of 26,000 (14/3/20) to maximum of 1,2500 (post operatively (28/10/19), WBC concentration ranging with minimum of 5000 to maximum of 25000. CRP was elevated to 30 mg/l (minimum)–107 mg/l (maximum). Liver function tests revealed total bilirubin of 0.43–0.6 mg/dl, alkaline phosphatase of 170–250 U/l, total protein of 4.5–7 g/dl, albumin of 2.8–3.5 gm/dl and globulin of 1.5–2.5 g/dl with A/G ratio of greater than 1.

ECG: NSR, Right axis Deviation, LV dominance (Figure 1).

CXR: Oligemic lung fields, small trachea, high branching of trachea, elevated right lower lobe, right lower lobe consolidation, cardiomegaly present.

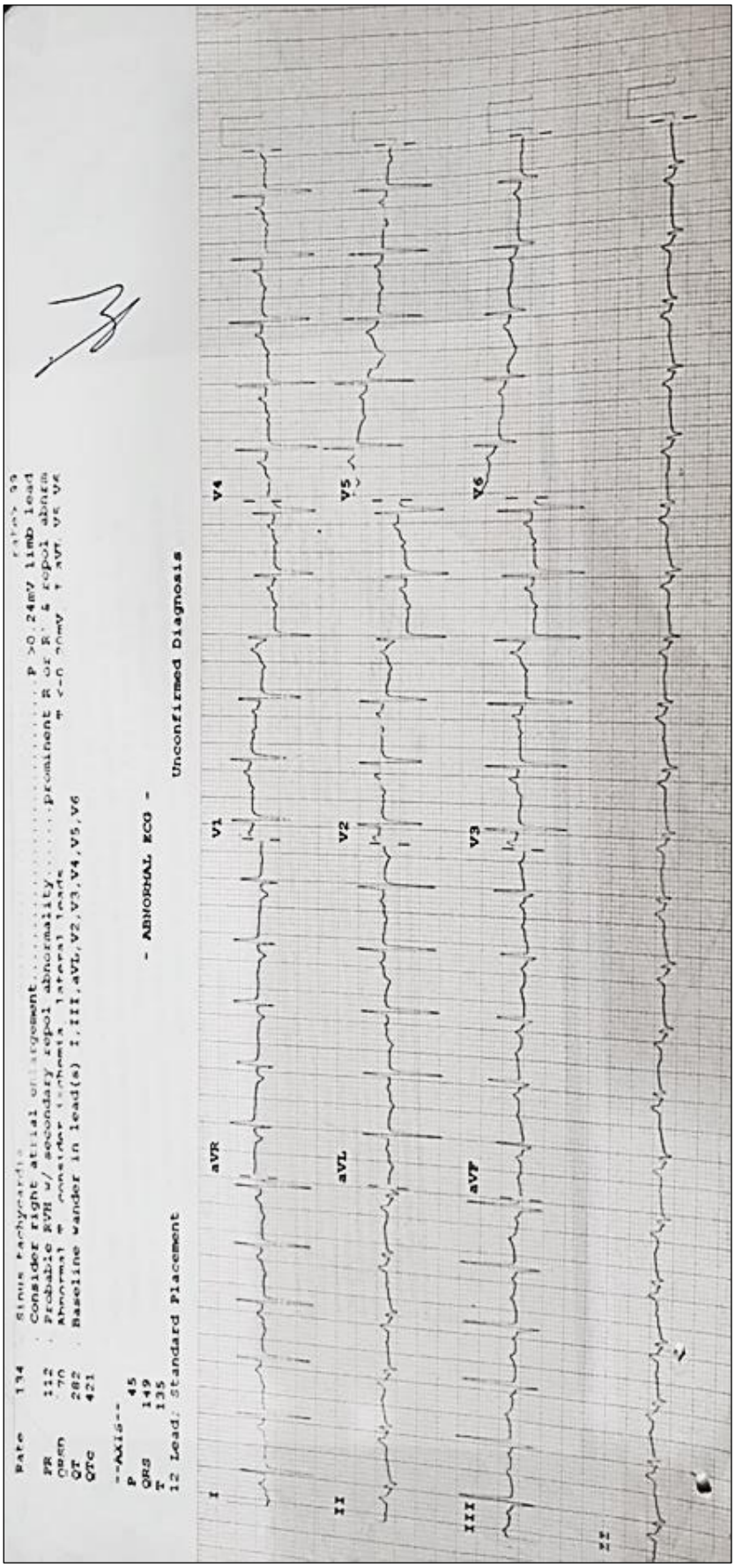

Fig. 1. *Electrocardiogram (ECG).*

Nutritional Requirements

Pre-Operative

Day 1: 13/3/2019 (NEC-1b)

Maintained via aminoven (1.5 ml/h) and IV fluids N/5D5% @3 ml/h.

Day 5: 19/3/2019 (NEC resolved)

Lactogen 10 ml Q3H, subsequently increased to 30 ml Q3H and then to 20 ml Q2H.

Post-Operative

2/5/2019 (3.6 kg)

Total no of feeds: 8 feeds/day; total volume: 160 ml.

EBM: 20 ml; sugar: 1 pinch; refined oil: 1 drop, providing total calories: 121 kcal/day, proteins: 1.76 g/day; protein/kg: 0.47 g/kg; calories/ml: 0.60 kcal.

23/7/2019 (no major improvement in weight)

Concentrate lactogen: 1 scoop in 25 ml (35 ml/feed),

Subsequently to be changed to dexolac: 1 scoop in 25 ml (35 ml/feed),

Addition of MCT oil: 0.5 ml/feed,

Vit. D drop: 1.5 ml/OD.

November–December: For proper weight gain and suspicion of GERD, child was kept on drip feeding by Ryles tube with no feeds at night from 12am to 5am (maximum weight of 5.9 kg in November).

26/11/2019 (5.2 kg)

Total volume of feeds: 550 ml, 11feeds/day,

Puffed rice feed: 30 ml (1/2teaspoon)*2 feeds; Ceralac feed: 30 ml (1/4 scoop)*2 feeds (with toned milk: 30 ml, sugar, refined oil).

Continuous pump feed: (from 4pm to 9 am): @35 ml/h (toned milk: 20 ml+ sugar (3 gm)+refined oil (1 ml),

Providing calories: 740 kcal/day; proteins: 14.9 g/day.

TREATMENT

The treatment of baby underwent various changes to counteract various difficulties during course of stay; various changes of antibiotics were made to counteract various challenges during course of stay.

Treatment on discharge:
Syp. Furoped 2 mg BD,
Tab. Aldactone 3.125 mg BD,
Syp. Levera 80 mg BD,
Syp. phenytoin 1.5 mg BD,
Syp. Osteocalcium 2.5 ml BD,
Syp. Lanzol 5 mg OD, and
Tab sildenafil 1 mg TDS.

LITERATURE REVIEW OF DISEASE CONDITION

Pentalogy of Fallot is a rare congenital heart defect comprising classic four features of Tetralogy of Fallot (VSD: Ventricular Septal Defect, Pulmonary Stenosis, Overriding Aorta and Right ventricular hypertrophy) with addition of an atrial septal defect (ASD) or patent ductus arteriosus (PDA).

ASD is abnormal opening between right and left atrium which allows blood to move from higher pressure left atrium to flow into lower pressure right atrium.

1. Ostium primum,
2. Ostium secundum,
3. Sinus venosus defect.

Patent ductus arteriosus is failure of fetal ductus arteriosus (artery connecting aorta and pulmonary artery) to close within first few weeks of life that allows blood to flow from higher pressure aorta to lower pressure pulmonary artery, which causes a left-to-right shunt [1].

Incidence

5 to 10% of all congenital heart diseases comprise of TOF with incidence of 3 in 10000 live births [2]. An additional ASD is seen in 3 to 5% of cases, most

commonly with ostium secundum type, followed by ostium primum type and rarely sinus venosus type [3]. Prevalence of congenital heart disease in India is approximately 4 cases per 1000 live births with TOF comprising 7–32% of these cases. There lies 40% chance of patients having associated cardiac conditions like anomalies of coronary arteries, right sided aortic arch, collateral vessels between systemic and pulmonary vasculatures, PDA, ASD and various aortic arch problems and regurgitations [4].

Clinical Manifestations

Some infants present acutely cyanotic at birth but others have mild cyanosis that progresses over first year of life as pulmonic stenosis worsens.

a. Characteristic systolic murmur.
b. Blue spells or tet spells: episodes are not seen in first couple of months of life, incidence peaks between second and fourth months after birth.
c. Anoxic spells during crying or after feeding (risk for emboli, seizures, and loss of consciousness or sudden death after an anoxic spell).
d. Poor feeding patterns with prolonged fussiness and agitation.
e. Squatting.

PDA C/M

a. Patients may be asymptomatic or show signs of HF.
b. Characteristic machinery-like murmur.
c. A widened pulse pressure and bounding pulses (due to overriding aorta).

ASD C/M

a. Patients may be asymptomatic.
b. Characteristic systolic murmur with a fixed split S2.
c. Diastolic murmur may be presented [5].

Case study child had tet spells (observed later in the course of the disease, not present at birth), poor feeding, anoxic spells, an increased pulse pressure murmur was not present due to the small size of the PDA.

Workup

Physical Examination

a. Clubbing of terminal digits (evidence of prolonged cyanosis).
b. Palpation: Medial displacement of Apical pulse (right ventricular enlargement).
c. Systolic thrill (palpable vibration felt over heart).
d. Auscultation: Systolic ejection murmur best heard along left sternal border (pulmonic valve region).
e. The second heart sound frequently is heard as single heart sound vice normal splitting found with the physiologic closure of aortic valve a few milliseconds before pulmonic valve, as a result of pulmonic valvular compromise or atresia.
f. Murmur is commonly not heard in patients with TOF due to enormity of defect and relative lack of turbulent flow.

Clubbing of the Digits and Medial Displacement of the Apical Pulse were Observed in the Baby.

ECG: Right axis deviation is usually noted due to these children commonly presenting with right ventricular hypertrophy [6].

Right Axis Deviation in Present in the ECG, LV Dominance is Also Seen.

Chest Radiography

a. Normal heart size.
b. Characteristic changes in shape: "BOOT shaped".
c. Lungs appear darker than normal.

The lung fields in the above case were oligemic suggesting features of TOF, though child did primarily not present with cardiomegaly, during the prolonged stay cardiomegaly was evident in the CXR with

other comorbid condition which included a small trachea, elevated right lower lobe with right lower lobe consolidation.

Imaging Studies: These tests are performed to identify TOF and associated anomalies that accompany TOF. These studies can also be useful in assessing the degree of severity of condition (echocardiography, CT and MRI scanning, and cardiac catheterization) [6].

SURGICAL MANAGEMENT

Surgery is majorly adopted intervention for management of congenital heart diseases. The approach can be palliative and corrective and complications are relatively common.

Corrective Surgical Interventions: closure of VSD using a patch. The infundibular obstruction can be surgically resected, with excessive tissue being sliced away to reduce the amount of restriction. Elective repair is usually performed in first year of life.

Complete repair involves closure of the VSD and resection of infundibular stenosis, with placement of a pericardial patch to enlarge the RVOT. Procedure requires a median sternotomy and use of cardiopulmonary bypass. Direct suture closure of associated ASD can be done. PDA clipping can be performed for PDA.

Palliative Surgical Interventions: Shunts to accommodate additional blood flow. The preferred procedure is a modified Blalock-Taussig shunt operation, which provides blood flow to pulmonary arteries from left or right subclavian artery via a tube graft. Shunts may result in pulmonary artery distortion [7].

The baby *underwent ICR+RVOT resection + Direct suture closure of ASD+PDA clipping on 28/3/2019.*

Prognosis

Operative mortality for total correction of tetralogy of Fallot is less than 3% (Jacobs, Mavroudis, Jacobs, and others, 2004). With improved surgical techniques, there is lower incidence of dysrhythmias and sudden death. Heart failure may occur postoperatively [8].

If untreated, TOF results in progressive right ventricular hypertrophy due to increased resistance on right ventricle. This progresses to heart failure which begins in right heart and often leads to left heart failure. Actual survival for untreated Tetralogy of Fallot is approximately 75% after first year of life, 60% by 4 years, 30% by 10 years, and 5% by 40 years. Severity of TOF is dependent on degree of right ventricular outflow tract obstruction. Only about 11% of patients with TOF will survive to 20 years of age without some form of intervention [9]. Approximately 96% of patients who do have surgical correction of this congenital heart defect are free of need for repeated surgical intervention for up to 20 years [10].

NURSING MANAGEMENT IN ICU

Short Term Goals	*Long Term Goals*
1. To improve cardiac function and decrease cardiac demands.	1. To encourage rehabilitation of child.
2. Improve tissue oxygenation and decreased oxygen consumption.	2. To prevent all complications.
3. To relieve respiratory distress.	3. To improve knowledge regarding home care and follow up.
4. To improve nutrition and activity tolerance.	
5. To maintain the skin integrity and prevent infections.	
6. To maintain fluid and electrolyte balance.	
7. To reduce fear and anxiety of parents.	

Assessment: Baby is Crying and Turns Cyanotic. Repeated Episodes are Presented. Symptoms Exaggerate at the Time of Feeding or Manhandling

Nursing Diagnosis

Activity intolerance related to hypoxia or decreased myocardial function.

Objective
To improve oxygenation and activity tolerance.

Interventions
a. Administration of titrated amount of oxygen to reach target oxygen saturations.
b. Assess response to oxygen therapy: increase in baseline oxygen saturations, improved work of breathing, and change in patient comfort.
c. Child is positioned so as to prevent aspiration of secretions.
d. Judicious use of suctioning procedure as needed.
e. For children who cannot take proper feeds, a nasotracheal or orotracheal tube is inserted to maintain proper feeds.
f. Signs of respiratory distress are monitored.
g. Blood gas analysis.
h. Chest physiotherapy.

Evaluation
Oxygenation is improved and baby had SpO2 between 80 and 90%.

Assessment: Child Appears Cyanotic in between, Perfusion is not Appropriate
Nursing Diagnosis
Decreased cardiac output related to decreased myocardial function.

Objective
To improve cardiac output.

Interventions
a. Remove accumulated fluid and sodium: Treatment consists of diuretics, possible fluid restriction and possible sodium restriction.
b. Organization of nursing care and medication schedule to provide periods of uninterrupted rest.
c. Decrease cardiac demands: Workload on heart is reduced when metabolic needs are kept to a minimum, accomplished by limiting physical activities, preserving body temperature, treating infection, reducing effort of breathing.
d. Improve tissue oxygenation and decrease oxygen consumption: Supplemental cool humidified oxygen is usually provided to increase amount of oxygen during inspiration.
e. Maintaining state of normothermia.
f. Monitored serum electrolyte levels and signs of hypotension.
g. Monitoring blood pressure and other vital signs every hour.

Evaluation
Peripheral perfusion is adequate.

Assessment: Risk for Infection Related to Chronic Illness
Nursing Diagnosis
Risk for infection related to chronic illness.

Objective
To prevent infection.

Interventions
a. Strict aseptic precautions for every invasive procedure.
b. Maintenance of neutral thermal environment.
c. Administered antibiotics as per order.
d. Maintenance of daily hygiene of baby and meticulous skin care.
e. Advice to parents about meticulous hand hygiene before touching child.
f. Avoided contact with infected patients.
g. Provided optimum nutritional status to support body's natural defenses.

Evaluation
The risk of infection has reduced significantly.

Assessment: Prolonged Hospitalization and Administration of Feed through Nasogastric Tube
Nursing Diagnosis
Altered nutrition related to reduced intake through NG tube with continuous ventilator support.

Objective

To meet the nutritional requirement of child and meet demands in accord with growing age.

Interventions

a. Nutrition is provided given by nasogastric or gastrostomy tube.
b. Nasogastric tube is usually taped in place with care to prevent pressure on the nares.
c. Most children have continuous feedings, but if bolus feedings are used, tube is rinsed with water after each feeding.
d. Skin and mucous membranes are examined for signs of dehydration.

Evaluation

Child's nutritional needs have been fulfilled.

HEALTH EDUCATION

Regarding Digoxin Administration

Explained parents how to take apical pulse and not to administer drug in case the apical pulse is less than 90 bpm.

Regarding Energy Conservation

Explained parents to provide a safe, calm environment and there should not be any extremes of environment which may cause fever in patient and bodily metabolic demands are increased.

Regarding Hygiene

- Explained parents about maintenance of hygiene in the hospital as well as home.
- Explained to keep the perineum and folds of skin clean and hand washing before and after doing any care of child.

Regarding Tube Feeds

Explained parents how to position baby after each feed so as to prevent aspirational pneumonia.

Regarding Position of Child

Told parents to maintain correct posture of child (semi prone or side lying position) to prevent aspiration and to elevate head end bed post feeds.

CONCLUSION

A variation of tetralogy of Fallot, pentalogy of Fallot is rather a rare congenital malformation with only 3–5% of the children with TOF. The definitive treatment includes of a surgical correction of the stenosed pulmonary arteries and patch closure of VSD and ASD. The case study in detail went through all the different approaches taken for the baby for holistic care perspective. The case study gives a detailed information regarding the surgical, nursing care, and nutritional care for the baby.

CONFLICT OF INTEREST

The author has no conflicts of interest.

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