

we think that infusion of neuromuscular blocker agents may provide better relaxation than fractional bolus ejection.

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Sickle cell disease in a woman with triplet pregnancy

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Triplet pregnancies are by nature rare, reported as 0.1% in a large series of cases.¹ Assisted reproductive technologies have multiplied their rate of occurrence in the last 2 decades. Whether spontaneous or induced, triplet pregnancies present with obstetrical and neonatal problems several fold more often than singleton or even twins. The rate of preterm birth among triplet pregnancies are reported as 96% and still birth rate 33.8%.² The major neonatal morbidity associated with triplet gestations is preterm delivery and low birth weight.³ Pregnancy in patients with hemoglobinopathy is associated with increased risk of maternal and perinatal morbidities and mortalities. Multiple pregnancy is potentially more hazardous than singleton pregnancy. There is a dearth of information concerning

multiple pregnancies in patients with hemoglobinopathy.⁴

We present a case of spontaneous triplet pregnancy in a patient with sickle cell disease. A 26-year-old Omani lady, gravida-2, para-1 was referred to the Obstetrics outpatient clinic of Sultan Qaboos University Hospital, Sultanate of Oman at 14 weeks of gestation. She has a known case of sickle cell disease and an ultra sonogram in a peripheral hospital at 9 weeks showed a triplet pregnancy. Her booking hemoglobin was 6.9 gm/dl and she received 2 units of packed red blood cell (PRBC) transfusion in the referring hospital. Her menstrual cycles were regular and she has not used any ovulation inducing agents or contraceptives. Her first pregnancy was also spontaneous, with a term delivery of a male baby weighing 2700 gm a year ago. She had postpartum hemorrhage requiring 2 units of PRBC. She had several admissions during that pregnancy for vaso-occlusive crisis or for blood transfusions. The patient was diagnosed to have sickle cell disease S/ thal since childhood, requiring several admissions to the hospital. Her 4 siblings are also suffering from the same disease. She had a non-consanguineous marriage and her husband is normal. Clinical examination revealed pallor and a uterine size corresponding to 24 weeks of gestation. Ultrasound examination showed 3 viable fetuses of 14 weeks gestation and a prophylactic cervical cerclage was inserted after a week. She continued to have regular follow up with hematologist and obstetrician and was put on oral penicillin V and folic acid supplements. Serial ultrasonogram showed satisfactory growth of all 3 fetuses. She required several admissions for top up and exchange transfusions. There was no evidence of any infection during pregnancy. At 26 weeks she was given 2 doses of injection dexamethasone to promote fetal lung maturity in case she goes into preterm labor. An elective cesarean section was planned at 32-34 weeks of gestation, but at 30 weeks she was admitted with labor pains. Cervical cerclage was removed and an emergency lower segment cesarean section was performed after arranging full neonatal support.

The details of the newborn babies are shown in **Table 1**. The placenta was trichorionic triamniotic weighing 1100gms. The estimated blood loss was 600 mls and she received 2 units of PRBC transfusion after surgery. Received 300 µgm of anti-D as she was Rh negative and without antibodies. Postoperative period was uneventful and she was discharged on the 6th day. At 6 weeks postpartum checkup she was clinically well, lactating and started on injection Depo-Provera for contraception. Babies at 3 months of age showed normal serial growth in all parameters and there was no evidence

of any retinopathy of prematurity. The incidence of multi-fetal gestation has increased significantly since the introduction of ovulation induction therapy and in-vitro fertilization and embryo transfer techniques. Multi-fetal gestation is associated with increased frequency of maternal complications and higher perinatal morbidity and mortality. As the number of fetus increases, the duration of gestation and birthweight decreases.⁵ The mean gestation at birth for triplets was reported as 33.5 ± 2.3 weeks and the mean birth weight as 1810 ± 270 gm for monochorionic pairs compared to 2125 ± 265 gm for dichorionic triplets.⁶ Our triplets were born earlier at 30 weeks and the mean birth weight was lower, only 1323 gm probably due to the sickle cell disease. Pregnancies complicated by sickle cell disease were significantly more likely to be associated with anemia, preterm delivery, proteinuric hypertension, birth weight below the 10th centile and emergency cesarean section. Severe sickling complications occurred more commonly in third trimester and there was some evidence that a prophylactic transfusion program reduced this risk. However, prophylactic transfusion did not improve obstetric outcome when compared with those pregnancies that were not transfused.⁷ Koshy et al⁸ did not find any significant difference in perinatal outcome between the offspring's of mothers with sickle cell disease who received prophylactic transfusion compared to those who did not receive. Prophylactic transfusion significantly reduced the incidence of painful crises of sickle cell disease but the increase in cost, number of hospitalizations and the risk of alloimmunization were disadvantages of prophylactic transfusion.⁸ Our patient was admitted several times with crises or anemia and had several

exchange and simple transfusions during pregnancy. We do not follow a policy of routine prophylactic transfusion but given for selected cases with previous poor obstetric outcome.

Meticulous care by obstetrician and hematologist with increased fetal surveillance and elective cesarean delivery are suggested in the management of such patients.⁴ Also, with the neonatal complications documented in these preterm babies, a highly functional neonatal intensive care unit is necessary.

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Table 1 - Details of the triplets.

Presentation	Gender	Weight	Apgar	Problems	Outcome
Cephalic	Girl	1400 gm	7 at 1 minute - 8 at 5 minute	RDS, metabolic acidosis. Ventilated x 15 hr.	Discharged on 35th day. Weight 1645 gm. At 40 weeks corrected age weight 2950 gm, height 45 cm.
Cephalic	Boy	1270 gm	7 at 1 minute - 8 at 5 minute	RDS, metabolic acidosis. Ventilated x 15 hr.	Discharged on 35th day. Weight 1820 gm. At 40 weeks corrected age weight 3180 gm, height 49 cm.
Cephalic	Girl	1300 gm	7 at 1 minute - 8 at 5 minute	RDS, metabolic acidosis. Ventilated x 15 hr.	Discharged on 35th day. Weight 1830 gm. At 40 weeks corrected age weight 3080 gm, height 48 cm.
RDS - respiratory distress syndrome.					

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Bronchiectasis following repair of esophageal atresia and tracheo-esophageal fistula

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The incidence of esophageal atresia and tracheoesophageal fistula (EA/TEF) was reported to be 1 in 4000-5000 live births.¹ Recurrent aspiration pneumonia is the most common complication described according to different mechanisms due to spill over of secretions through TEF or esophageal pouch, esophageal dysmotility, gastro-esophageal reflux (GER) and absence of ciliated epithelium in the trachea.² Many other causes of respiratory complications including esophageal dysmotility, tracheomalacia, anastomotic stricture, and recurrent or double fistula were described previously.^{3,4} In this report, we describe bronchiectasis as a complication following repair of EA and TEF. We undertook a retrospective review of the charts for all EA/TEF patients referred to the pulmonary clinic for evaluation of recurrent chest infection and preoperative evaluation during the period from November 1993 to October 2004 at the main tertiary care center for referral of complicated cases in Saudi Arabia. Bronchiectasis was diagnosed based on dilated bronchi on CT chest.

There were a total of 41 patients, 26 (63%) males and 15 (37%) females. Forty patients (98%) are alive and one (2%) died. Fourteen (34%) were premature and 27(66%) were full term. The TEF was diagnosed at birth in 34 (83%) of the patients. Patients were referred to King Faisal Specialist Hospital and Research Center at 15±29 months. The period of follow up was 5±3.8 years. Diagnosis of TEF was based on nasogastric tube coiling (NGT) and by dilated blind esophagus on chest x-ray in 40 (98%) of the patients. An EA and distal TEF were found in 37 (90%) of the patients, isolated EA in 2 (5%) and H-type fistula in 2 (5%) of the patients. Congenital anomalies were associated in 28 (68%)

of the patients. Cardiac anomalies were found in 11 (27%), gastrointestinal (GIT) in 8 (20%), respiratory system anomalies in 12 (30%), renal in 7 (17%), skeletal in 12 (30%), and chromosomal in 7 (17%). More than 1/3 of the patients had post-operative complications including pneumothorax, recurrent fistula, leakage at operation site and empyema. Thirty (73%) presented with pneumonia and required prolonged ventilation. Esophageal dysmotility and GER developed in >90% of the patients. Twenty-four (60%) of the patients required Nissen fundal plication for GER. Esophageal stricture that required >3 dilatations developed in 16 (46%) of the patients. The GER was significantly related to development of atelectasis, dysmotility, and aspiration pneumonia ($p<0.05$), but not related to surgery type if it is primary anastomosis or staged surgery ($p>0.05$). Pulmonary complications developed in >70% of the patients including persistent atelectasis, chronic aspiration pneumonia, asthma or hyper reactive airway disease, and chronic lung disease that required oxygen for more than one month. Tracheomalacia occurred in 12 (29%) of the patients. Bronchiectasis developed in 7 (17%) of the patients (Table 1), 2 of them after gastric tube replacement of esophagus, one after colonic replacement, and 4 developed after primary repair. Two of the 4 patients with primary repair were premature, another one with multiple congenital anomalies and the 4th one with recurrent fistula, esophageal diverticulum and cardiac anomalies (Table 1). Pulmonary function test (PFT) was carried out in 16 (40%) patients who were able to comprehend the test maneuver. Eighty-eight percent of patients who performed PFT showed abnormal values: obstructive PFT changes in 3 (7%), restrictive in 8 (20%), combined obstructive and restrictive changes in 3 (7%) and normal in 2(4%).

Long-term pulmonary complications have been described before.^{2,4-6} Couriel et al⁴ described bronchitis for more than 8 years in 5/20 patients (25%), and denoted that lung disease improves with time. Chetcutti et al⁵ described asthma development in 40/155 (26%) patients after TEF repair, with restrictive lung changes in 18 (12%) of the population. Delius et al² showed that 31/68 patients (46%) developed recurrent pneumonia that required 1-10 admissions to hospital for treatment. Robertson et al⁶ performed PFT in 25 patients with TEF repair and their siblings and found that, although PFT values were within normal limits, they were significantly different compared to their siblings. The later study also showed that 6/25 patients had positive methacholine challenge test as a sign of obstructive airway disease and 9/25 had a restrictive pattern. Gastroesophageal reflux as the primary cause of respiratory symptoms in these patients has