

genetically predisposed individuals. The mean time taken for defervescence in our patients was 34.2 days after starting anti TB drugs; this increased the length of hospital stay and caused additional costs to the health care system. These findings further emphasize the need for early detection, and treatment of MTB infection. More than 80% of our patients lost 20% or more of their body weight at the time of presentation and all of them were non-reactive to TST. These findings are in agreement with the previous results of Pilheu et al.,<sup>3</sup> who noted 20% or more of body weight reduction at the time of presentation, in a study of 17 patients with severe pulmonary TB. Our results are also in agreement with the results of Kony et al.,<sup>2</sup> reporting that patients with CD4+ T-cells count <300 x 10<sup>6</sup>/L are less likely to react to TST, and have higher frequency of extra-pulmonary involvement, in the form of pleural effusion, lymphadenopathy, miliary disease, and oral candidiasis.

In conclusion, the results of our study show that, active MTB infection can induce profound CD4+ T-lymphopenia, as seen in patients with HIV infection, however, the defect is reversible with the effective treatment of MTB infection. Further, studies are probably needed with a larger patient number to develop general guidelines for the management of this sub-group of patients, having this grave combination of active MTB infection, and CD4+ T-lymphopenia. It is worth reminding that, in patients found to have lymphopenia with MTB infection; one should carefully investigate for the presence of disseminated disease and check their CD4+ T-cell count. Patients with counts <300 x 10<sup>6</sup>/L have poor prognosis, particularly those with CD4+ T-cell count <150 x 10<sup>6</sup>/L have the highest mortality. Such patients should be managed in the setting of intensive care unit during the initial phase of their treatment.

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## Management of difficult airway in a child with arthrogryposis multiplex congenita during general anesthesia

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Arthrogryposis multiplex congenita (AMC) consists of complex congenital anomalies characterized with multiple contractures. The possibility of autosomal dominant inheritance with reduced penetrance is suggested for this apparently new syndrome.<sup>1</sup> This syndrome may include foot deformities such as pes equinovarus (PEV), and congenital convex pes valgus. There may be a variety of deformities of the knee: flexion deformity, genu recurvatum, and genu valgum. Hip deformities such as unilateral or bilateral dislocation contractures may also exist. Craniofacial abnormalities, multiple joint contractures, pulmonary hypoplasia, cryptorchidism, and unusual ophthalmological findings are the other characteristics of this syndrome. In some severely affected persons, the central nervous system may also be affected.<sup>2</sup> Arthrogryposis multiplex congenita, which is diagnosed at birth presents with multiple joint contractures. Anesthetic management of these patients requires special care. As this disease often progresses until dysfunction of multiple organ systems occur, it may have an impact on the anesthetic management. Difficult tracheal intubation may be encountered due to limited neck extension, inadequate mouth opening, and short epiglottis.<sup>3</sup>

A 3-year-old and 20 kg weight boy who was diagnosed as PEV was prepared for surgical correction of the deformity. During preoperative examination, there was no history of any congenital anomaly or marriage of close relationship. The other 2 brothers were healthy. His mother had history of non-specific febrile infection and had no treatment

in the early period of her pregnancy. The child had limitation in range of motions of his ankle, neck, hip, and knee since the neonatal period. We determined pale skin, dimorphic appearance of face, small and firm chin, stiffness, and limitation of range of motion of cervical spine, contractures in lower extremities, and difficulties in walking in physical examination. Any pathology of respiratory and cardiovascular system has not been established. We interpreted biochemical study, cranial CT, cranial MRI, and electroencephalogram as normal. Before the operation 0.2 mg/kg midazolam diluted with serum physiologic was given by rectal route. Intravenous injection (IV) route was established with 500 cc isolex P using 22-G catheter on left hand dorsal side 30 minutes after premedication. He was monitored to perform electrocardiogram (ECG), peripheral oxygen saturation (SpO<sub>2</sub>), noninvasive blood pressure (NIBP), end-tidal carbon-dioxide (EtCO<sub>2</sub>) and temperature in operating theater were in normal levels. For induction of anesthesia 10 µg/kg atropine, 3.0-mg/kg propofol, 2.0 µg/kg fentanyl, and 0.1 mg/kg vecuronium bromure were given via IV route and ventilation of the patient was supplied with oxygen 100% using facemask. Two minutes after administration of muscle relaxant, laryngoscopy was performed for intubation, but the he could not be intubated as epiglottis and vocal cords were not visualized. Ventilation of the patient was continued, and intubation manipulation was repeated. For intubation a fiberoptic laryngoscope, which has a blade that has an extension angle of 70° was used (Flexotype, Heine, Germany). He was intubated by the help of a guide inserted into a disposable tube of 4.5 ID diameter, and connected to an automated ventilator after setting tidal volume as 8 ml/kg (Cicero EM, Drager/Germany). Anesthesia was maintained with sevoflurane 2%, and nitrous oxide 60% in oxygen. The airway pressure, respiratory rate, tidal volume, EtCO<sub>2</sub>, and anesthetic gases were monitored and EtCO<sub>2</sub> was normal after intubation. The EtCO<sub>2</sub> at remained 40-53 mm Hg during operation. The airway pressure of the patient began to increase following intubation, after 30 minutes airway pressure exceeded 20 cm H<sub>2</sub>O, and vecuronium bromure 0.3 mg/kg was administered in bolus. The airway pressure did not decrease although the dose of muscle relaxant was repeated every 30 minutes throughout the operation, which lasted 120 minutes. He remained stable hemodynamically during the preoperative period. At the end of the operation when he entered into the awakening period, his muscle power did not improve, and atropine 10 µg/kg, and neostigmin 20 µg/kg were given in bolus for recurarisation. He was taken to the recovery room after the values of SpO<sub>2</sub> were normalized. He was observed until he was hemodynamically stable.

Arthrogryposis multiplex congenita is a congenital disorder that consists of multiple orthopedic anomalies. They are the candidates for difficult intubation due to small chin, stiffness of temporomandibular joint, and limitation of range of motion of cervical spine.<sup>3</sup> In the literature, 10 patients have been reported to present difficulty for their airway control related to AMC. Therefore, we should prefer fiberoptic laryngoscope, with a blade that has an extension angle of 70 degrees. Vocal cords can be visualized better and with the help of this type of laryngoscope can easily make intubation. The surgical manipulation would not be comfortable due to an inadequate muscle relaxation in the surgical areas. This situation may possibly be due to out of junction receptors. We think that infusion of neuromuscular blocker agents may provide better relaxation with 0.2 mg/kg/h than repeated fractional bolus ejection at 30-minute times. Therefore, regional anesthesia can be safely used for different surgical procedures in patients with AMC in the presence of appropriate monitoring.<sup>4</sup> When volatile anesthetic agents are used, the incidence of malignant hyperthermia is high due to scoliosis, abnormal subcutaneous tissue, and myopathic muscles. Up to now, 4 patients with AMC was reported to have hyperthermia. Furthermore, these patients may have accompanying congenital cardiac pathology in the rate of 10%, and this condition is very important in the anesthesia.<sup>3,4</sup> Hopkins et al, reported that 2 patients with AMC developed hypermetabolic reactions during anesthesia and surgery and they proposed that the reaction is distinct from malignant hyperthermia and independent of the anesthetic agents used.<sup>5</sup> It is suggested in the anesthetic management with AMC, low-dose ketamine be administered by continuous infusion, with a satisfactory result due to myopathic features, hypoplastic musculature, severe scoliosis, and expected increased sensitivity to various sedatives and volatile anesthetic agents. However, propofol can be a reliable agent for anesthesia in AMC patients who had hyperthermia and difficult intubation in previous surgical procedures.<sup>5</sup>

In conclusion, AMC, which is commonly associated with other diseases characterized with accumulation of abnormal material in the joint tissues, may cause increasingly severe airway problems with age. Anticipating and preparing for a difficult airway is probably more important than using any particular anesthetic technique. As vocal cords can be well visualized and intubation can easily be made with the help of fiberoptic laryngoscope, which has a blade that has an extension angle of 70°, this type of laryngoscope should be preferred. However, it is recommended that the facilities for fiberoptic intubation and an experienced pediatric anesthetist are present for all such cases. In addition,

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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**S** triplet pregnancies are by nature rare, reported as 0.1% in a large series of cases.<sup>1</sup> 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%.<sup>2</sup> The major neonatal morbidity associated with triplet gestations is preterm delivery and low birth weight.<sup>3</sup> 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.<sup>4</sup>

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