

## Our successful regional anesthesia experience in a patient with Kabuki Syndrome

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**K**abuki syndrome (KS) (Kabuki make-up or Niikawa-Kuroki syndrome) is a rare condition coursing with congenital malformation and mental retardation. "Kabuki make-up syndrome" name was coined by Niikawa et al. since the faces of the children exhibiting the syndrome resembles the makeup of the actors in traditional Japanese theater. In this report, our successful regional anesthesia experience with a 19 year old male KS patient who underwent operation due to patella dislocation is presented.

**Case:** An 19 year old male patient was followed up with the diagnosis of Kabuki makeup syndrome from birth on and orthopedics clinic which he referred to with the complaint of patella dislocation planned operation. Following routine monitorization, patients was sedated with 3 mg dormicum and, in consideration of the difficulty of intubation and the risk of malignant hyperthermia, femoral sciatic block was planned and was carried out with Braun 22g soft block needle accompanied by nerve stimulator.

For femoral-sciatic nerve block, 20 ml 0.5% bupivacain and 20 ml 2% prilocain mixture was prepared (overall 40 ml) and separate 20 ml injections were made to femoral and sciatic nerves. And patella osteotomy was carried out. No perioperative complication occurred.

**Discussion:** KS is a rarely occurring condition whose etiology remains unknown and which courses with multiple congenital malformation, and mental retardation. Abnormalities in KS, which are thought to influence anesthesia management, are micrognathia and cleft and high palate. In some patients, obesity and obstructive sleep apneas may emerge in adolescence due to hypotonia. Scoliosis occurring in 35% of the patients may influence respiratory function. Although muscular biopsies are normal, the presence of muscular hypotonia in these patients, and prolongation of neuromuscular blockage may decrease the risk of malignant hyperthermia

**Conclusion:** In conclusion, As KS is a rare occurrence, it is not usually considered and its diagnostic criteria apart from facial anomaly have not been established definitively, it is a syndrome which requires surgical procedures and hence anesthesia more and more due to anomalies. It is our belief that anesthetists should evaluate these patients thoroughly prior to operation, should take all measures against probable difficulty in intubation, and extubation, and the risk of malignant hyperthermia and take care against the risk of probable systemic and neurological complications in peri/postoperative period.

### Biography

I. Ozkan Onal has completed his medical education from Ankara Gazi University Medical School and he has completed his anesthesia training from Ankara Hacettepe University Medical School and he is working in Ankara Yuksek Ihtisas training and educational hospital. He has published more than 15 papers about anesthesia.

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