

Cellular and molecular medicine: its role in management of complicated uncommon disorders during anesthesia

In this issue of the Journal, two manuscripts are published that discuss two relatively uncommon diseases that anesthesiologists are often encountered when anesthetizing pediatric patients though these patients may be seen during adulthood. Caplan et al have introduced a and discussed anesthetic management of such patients a 19 month old Babyboy with history of Medium-chain acyl-coenzyme A dehydrogenase deficiency (MCADD) undergoing orchidopexy for undescended testis; the patient had been diagnosed during newborn screening assessment (1). In another case series, Mottaghi et al have discussed 3 patients with Klippel–Feil syndrome, spanning 6 to 42 years old patients having underlying disorder with complicated airway management (2). Both of these articles discuss challenges for anesthesiologists that mandate more sophisticated view towards cellular and molecular mechanisms of diseases affecting perioperative anesthetic management. In another article, Talebi et al have done a review on the cellular and molecular mechanisms in perioperative hepatic protection including a thorough evaluation of the mechanisms, drugs and methods of protection (3).

MCADD is known as "the most common disorder of mitochondrial fatty acid oxidation" as discussed by Wiles et al (4, 5), with involvement of many vital organs. In their conclusion for MCADD patients, Caplan et al have presented a few suggestions; their recommendations are in concordance with similar studies including Allen et al (6-8):

- Anesthetic management with special approach to multidisciplinary preoperative planning
- Avoiding prolonged fasting through a number of strategies including preoperative administration of dextrose added intravenous fluids and scheduling the patients as the first case of the list

- Logic and cautious use of anesthetic drugs including volatile anesthetics, neuromuscular blocking agents, and propofol

Klippel-Feil syndrome is known as a rare disorder involving mainly the cervical spine with abnormalities in fusion of at least 2 vertebrae; recent developments in cellular medicine has significantly affected our clinical practice in these patients (9-11). In their case series, Mottaghi et al have discussed each patient with its radiologic studies and finally, have concluded that in Klippel–Feil syndrome, the following strategies should be used; these considerations are discussed with some different viewpoints in other studies (2, 12, 13):

- Adequate assessment of the airway with fully prepared airway management arsenal
- Using awake fiberoptic intubation approach and if not feasible, using inhalational induction and fiberoptic intubation as an alternative

Talebi et al have discussed the very direct effects of cellular and molecular medicine on our daily anesthesia practice; with especial focus on risk factors for anesthetic drugs and the perioperative period events (3).

These findings once again notify us that cellular and molecular mechanisms of diseases directly affect our clinical perioperative practice. Based on these studies and many other recent studies, adopting interdisciplinary approach in anesthesia management and perioperative care is a necessity if we want to improve the quality of care in our patients (14).

References

1. Caplan LA FM. Anesthetic considerations in medium-chain acyl-CoA dehydrogenase deficiency. *J Cell Mol Anesth.* 2017;2(2):69-76.
2. Mottaghi K SF, Sezari P, Gholizadeh N, Nashibi M. Anesthetic

- management of patients with Klippel-Feil syndrome, a case series. *J Cell Mol Anesth.* 2017;2(2):63-8.
3. Talebi Z PH, Dabbagh A. Cellular and molecular mechanisms in perioperative hepatic protection: a review of current interventions. *J Cell Mol Anesth.* 2017;2(2):82-93.
 4. Wiles JR, Leslie N, Knilans TK, Akinbi H. Prolonged QTc interval in association with medium-chain acyl-coenzyme A dehydrogenase deficiency. *Pediatrics.* 2014;133(6):e1781-6.
 5. Schatz UA, Ensenauer R. The clinical manifestation of MCAD deficiency: challenges towards adulthood in the screened population. *Journal of inherited metabolic disease.* 2010;33(5):513-20.
 6. Redshaw C, Stewart C. Anesthetic agents in patients with very long-chain acyl-coenzyme A dehydrogenase deficiency: a literature review. *Paediatr Anaesth.* 2014;24(11):1115-9.
 7. Justiz AC, Mayhew JF. Anesthesia in a child with medium-chain Acyl-CoA dehydrogenase deficiency. *Paediatr Anaesth.* 2006;16(12):1293-4.
 8. Allen C, Perkins R, Schwahn B. A retrospective review of anesthesia and perioperative care in children with medium-chain acyl-CoA dehydrogenase deficiency. *Paediatr Anaesth.* 2017;27(1):60-5.
 9. Saker E, Loukas M, Oskouian RJ, Tubbs RS. The intriguing history of vertebral fusion anomalies: the Klippel-Feil syndrome. *Child's nervous system : ChNS : official journal of the International Society for Pediatric Neurosurgery.* 2016;32(9):1599-602.
 10. Kim HJ. Cervical spine anomalies in children and adolescents. *Curr Opin Pediatr.* 2013;25(1):72-7.
 11. Can A, Dos Santos Rubio EJ, Jasperse B, Verdijk RM, Harhangi BS. Spinal Neurenteric Cyst in Association with Klippel-Feil Syndrome: Case Report and Literature Review. *World neurosurgery.* 2015;84(2):592.e9-14.
 12. Spond M, Burns T, Rosenbaum T, Lienhart K. Crisis Management of Accidental Extubation in a Prone-Positioned Patient with Klippel-Feil Syndrome. *A A Case Rep.* 2016;6(12):383-6.
 13. Bakan M, Umutoglu T, Zengin SU, Topuz U. The success of direct laryngoscopy in children with Klippel-Feil Syndrome. *Minerva anesthesiologica.* 2015;81(12):1384-6.
 14. Rajaei S DA. Interdisciplinary Approach and Anesthesiology: Is There Any Role? *J Cell Mol Anesth.* 2016;1(3):129-33.

Ali Dabbagh
Professor, Fellowship in Cardiac Anesthesiology
Chairman and Editor in Chief
JCMA
Anesthesiology Research Center,
Shahid Beheshti University of Medical Sciences,
Tehran,
Iran