

[GA1-49] Anesthetic management of posterior spinal fusion in a patient with Ehlers-Danlos type VIIC (Dermatosparaxis)

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Introduction: Ehlers-Danlos type VIIC (dermatosparaxis) is a rare (7 cases described) autosomal recessive connective tissue disorder caused by procollagen 1 N-proteinase deficiency. Phenotypic features include extreme skin laxity, fragility and easy bruising. We report the anesthetic management of a patient undergoing extensive spinal fusion.

Case Report: A 24 year old, 49 kg female with severe back pain secondary to thoracolumbar kyphoscoliosis presented for posterior instrumented spinal fusion from T9 to her pelvis. Anesthetic management included PICC line placement prior to induction. After easy intubation, the endotracheal tube was wired to her premolars to avoid facial tape. Even careful laryngoscopy produced a pinpoint skin tear through her lower lip which rapidly propagated through her entire lip. Because of redundant skin on her distal extremities, a 20 gauge angiocatheter was placed in her basilic vein which infiltrated 4 hours into her procedure. Sequentially placed 22 g. bilateral radial arterial lines were easily placed but failed rapidly after the formation of large forearm hematomas. Noninvasive blood pressure monitoring was performed infrequently due to formation of edema and ecchymosis at the cuff site. Her head was placed in Mayfield tongs to prevent pressure on facial tissues and pressure points were padded with silicone gel sheeting, Mepilex Border Lite dressings and Allevyn foam. After she was gently elevated and manually log-rolled by multiple assistants onto the Wilson frame (mounted on a Jackson table) she developed a 10 inch complete skin tear on her chest. SSEP monitoring was unsuccessful and MEPs were not used because of an inability to secure bite blocks adequately. A successful wake-up test was performed after orthopedic fixation. Final repositioning, without trauma, to supine was accomplished by firmly mounting and locking the padded Jackson table to the patient's back to sandwich the patient and rotating axially 180 degrees to supine position. The patient was extubated and admitted to the ICU.

Discussion: There are multiple anesthetic concerns for the dermatosparaxis patient who presents for a major surgery. This report highlights the difficulties in obtaining and maintaining venous and arterial access. Invasive blood pressure monitoring may be preferred as tissue trauma may occur with serial noninvasive measurements. If arterial cannulation is attempted, Seldinger technique may be superior to transfixion, potentially causing less trauma to lax tissues. If spine surgery is planned, standard neuromonitoring may be compromised. Additionally, prone positioning via axial rotation of the Jackson table may place less tension on fragile skin than a log-rolling method. Padding of pressure points is of paramount importance.

Picture shown is an example of the patient's lax and redundant skin:

References:

1. De Paepe A et al. Clin Genet 2012; 82: 1–11.
2. Malfait F et al. Am J Med Genet A 2004; 131 (1): 18–28.

