

## **Spondylo-Epiphyseal dysplasia - A challenge for operative positioning**

Sir,

Spondyloepiphyseal dysplasia tarda (SDT) is a rare autosomal recessive disorder of bone growth.

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These patients are likely to have kyphoscoliosis, platyspondyly, pectus carinatum and clubfoot. They may also develop arthropathy of large joints.<sup>[1]</sup> These deformities may lead to impediment in safe positioning during surgery, which subsequently may lead to various neurovascular complications. We report a patient with SDT who developed venous congestion of the left upper limb owing to arthropathy of elbow joint following a spinal surgery in prone position.

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**Figure 1:** Patient with spondylo-epiphyseal dysplasia tarda features and bilateral elbow arthropathy with flexion deformity

A 19-year-old male with SDT presented with backache and bilateral lower limb weakness with bowel-bladder involvement. He had short stature with short neck, arthropathy of large joints, clubfeet and fixed flexion deformities at elbow and knee joints [Figure 1]. Imaging revealed multi-level platyspondyly with reduced canal diameters of cervical and dorsal spine, cord edema and dysplastic changes in the heads and distal ends of femur and humerus. The patient was scheduled for posterior decompression and instrumentation from C5 to T10 vertebral levels. The patient was shifted to operating room and general anaesthesia was induced. Intubation was attempted using video-laryngoscope with in-line stabilisation (fiberoptic bronchoscope was not available). Three attempts were taken; however, no part of glottis was visualised. Any further attempts were not tried due to possibility of atlanto-axial instability.

Pro-seal laryngeal mask airway (LMA) was inserted and tracheostomy was done with minimal neck extension. Right radial artery was cannulated for invasive blood pressure monitoring. The patient was positioned prone on Jackson Table. During positioning, arthopathic joints were carefully padded with cotton. The optimal positioning of the patient was cumbersome. Following multiple attempts, near-optimal position was achieved. Following the final positioning, peripheral arterial pulsations were also checked in all four limbs and were equally present. Surgery lasted for approximately 9 hours, during which 1500 ml of blood loss occurred. Intraoperative vitals remained stable. At the end of surgery patient was turned supine, residual paralysis was reversed. A bluish discoloration with swelling of the left forearm was immediately noted [Figure 2]. It was not associated with pain on rest



**Figure 2:** Venous congestion of left forearm postoperatively

or at movement of limb. Pulsations of radial as well as ulnar arteries in the left side were well palpable. A clinical diagnosis of venous obstruction leading to venous stasis was made. The cause was diagnosed as a widened humeral distal metaphysis, which might have led to obstruction to the venous drainage of the forearm in the prone position. A magnesium sulfate dressing was applied and limb was elevated. Arterial pulsations were checked hourly and patient was asked to mobilise the fingers frequently. After few hours, the discoloration disappeared. The spasticity of both the lower limbs improved following surgery. The patient was uneventfully discharged 8 days later.

Presence of multiple skeletal deformities in patients, pose problems for positioning.<sup>[2,3]</sup> Prone position is known to predispose venous stasis.<sup>[4]</sup> In this patient, the combination of reduced venous return owing to prone positioning and elbow joint arthropathy may have led to the venous outflow obstruction of forearm and hand. Pulse oximetry, which was placed on the left thumb, could not have detected this venous obstruction, as it detects poor perfusion secondary to arterial occlusion. Use of frequent non invasive blood pressure monitoring should be avoided in the patients with such deformed joints.

Venous occlusion may lead to reduced microvascular flow and resultant tissue hypoxia. There are various experimental modalities to continuously measure peripheral venous oxygenation such as use of near-infrared spectroscopy-venous occlusion technique (NIRS-VOT), using photo-plethysmograph (PPG) signals and venous oximetry technique using

artificially generated venous pulsation.<sup>[5,6]</sup> Studies have utilised NIRS-VOT to detect reduced muscle flows and hypoxia due to venous occlusion.<sup>[5]</sup> Once such monitors are commercially available, these can be a boon to monitor such patients and prevent such complications.

We conclude that, in patients with skeletal dysplasia and arthropathies posted for surgeries, extra vigilance such as intermittent examination and intermittent repositioning of limbs is needed to prevent positioning related neurovascular compromise even for short duration of surgery or surgery in supine position.

#### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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#### Conflicts of interest

There are no conflicts of interest.

**Konish Biswas, Priyanka Gupta, Ashutosh Kaushal, Iftthekar Syed<sup>1</sup>**

Departments of Anaesthesia and <sup>1</sup>Orthopaedics, AIIMS, Rishikesh, Uttarakhand, India

**Address for correspondence:**  
Dr. Priyanka Gupta,

Department of Anaesthesia, AIIMS, Rishikesh, Uttarakhand, India.

E-mail: drpriyankagupta84@gmail.com

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