Ultrasound-assisted spinal anaesthesia in a patient with Wildervanck syndrome and congenital abnormalities of the lumbar spine

Editor—Wildervanck syndrome is a rare condition characterized by Klippel–Feil (KF) deformity of the cervical spine, Duane’s retraction syndrome (abducent’s palsy with retracted bulbi), and hearing loss.1 The main consideration of the KF deformity is a difficult airway. Regional anaesthesia has been recommended as an approach to avoiding the challenges posed by a difficult airway, but there have been no reports of spinal anaesthesia in patients with this condition.2

Our patient was a 61-yr-old female who presented for resection of a soft tissue tumour in her left lower leg. She gave a history of Wildervanck syndrome, which consisted of the absence of the atlas and fusion of C2–3, resulting in markedly reduced cervical spine range of motion, multiple predictors of difficult laryngoscopy, and fusion of the facet joints in the lumbar spine. Her recent anaesthetic history included an awake fibreoptic intubation and general anaesthesia for laparoscopic cholecystectomy. Her medical history included obstructive sleep apnoea, essential hypertension, and a BMI of 41.

The patient was brought to the operating theatre, where spinal anaesthesia proved to be challenging. Eventually, after several attempts, a 22 G Quincke needle was successfully placed at L3–4 via a midline approach after multiple needle redirections in the caudal-cephalad and medial-lateral directions met with bony resistance or paraesthesia.

Twelve weeks later, the patient required further surgery on the same leg. On this occasion, an expert in regional anaesthesia and ultrasound techniques performed a pre-procedural ultrasound surveillance of the lumbar spine. This demonstrated extensive fusion of lumbar facet joints and narrowing of the intervertebral and interlaminar spaces except at L3–4 (Fig. 1) where a 22 G Quincke needle was inserted successfully.

This report highlights a case of Wildervanck syndrome where spinal anaesthesia proved to be difficult due to lumbar spine abnormalities but was easily achieved on a second occasion with ultrasound assistance. This finding has not been reported before. The best known anaesthetic implication of Wildervanck’s syndrome is the airway as a consequence of the KF anomaly.1–3 Patients with KF syndrome have a variable degree of cervical fusion and congenital malformation/absence/fusion of the cervical vertebrae. As a consequence, the neck is webbed, short, thick, and immobile, appearing to sit directly on the trunk.5 Neck movements may be restricted secondary to absent or fused cervical segments.3 In keeping with the oculo-auriculo-vertebral spectrum of congenital anomalies, patients with Wildervanck syndrome may exhibit hypoplasia of the molar, maxillary, or mandibular region, asymmetric facies, torticollis, and cleft lip and/or palate.6–7 These features may compromise bag-mask ventilation. Patients with Wildervanck’s syndrome should be thoroughly assessed clinically and radiologically for C-spine abnormalities with plain radiographs or computed tomography.6 If airway management is required, awake fibreoptic intubation represents the safest approach. C-spine precautions should be respected and inline stabilization performed during direct laryngoscopy and intubation.5

Neuraxial techniques where appropriate are recommended so as to avoid airway manipulation.2 However, it is not widely known that lumbar and thoracic spine abnormalities may be present. These are in keeping with a type III KF anomaly seen in Wildervanck and represent a more severe form of KF.7 A detailed history to identify back problems accompanied by ultrasonographic imaging of the lumbar spine

Fig 1 Transverse view at L3/4. Laminae indicated by * and transverse processes indicated by +.
are recommended when neuraxial techniques are planned. Ultrasound-assisted spinal anaesthesia may be helpful with challenging spinal anatomy but is not always successful and real-time ultrasound-guided spinal anaesthesia may be necessary.\textsuperscript{8} However, an experienced practitioner in ultrasound-guided spinal techniques may be required.

Declaration of interest
None declared.

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Outcomes and patterns of infections in patients with underlying haematological malignancies admitted to intensive care

Editor—We read with interest the article by Bird and colleagues\textsuperscript{1} ‘Outcomes and prognostic factors in patients with haematological malignancy admitted to a specialist cancer intensive care unit: a 5 yr study’. Patients with haematological malignancies admitted to an intensive care unit (ICU) with an acute illness have experienced significantly improved outcome in recent years, in comparison with observed hospital mortality rates of up to 80% in the 1980s in those developing respiratory failure.\textsuperscript{2} The current paper provides further credence to this observation reporting spectacular survival rates in acutely ill haematology patients admitted to ICU.\textsuperscript{3} Despite significant improvement in the last few decades, high risk of death still exists in those requiring multiple organ support, and those who do survive have a long recovery time, including prolonged ICU and hospital lengths of stay (LOS).

In a retrospective and parallel study recently conducted in our own ICU in a tertiary referral centre for haematological diseases in London, UK, we audited outcomes of acutely ill haematology patients admitted to our ICU. The audit was part of a service evaluation for which local R&D approval was obtained. In addition to survival outcome, we extended our query to study the patterns of infections seen in acutely ill haematology patients in our ICU that is unlike previously published studies, which studied infections in a non-critically ill haematology patient population.

All consecutive admissions of adult patients to the ICU with an underlying haematological malignancy between January 2005 and December 2008 were included. The data collection involved information obtained from the ICU and the Pathology databases, with a senior haematologist independently verifying the haematological details.

Ninety-seven out of the total 3880 (3%) patients admitted to ICU during the study period had an identifiable underlying haematological malignancy. Fifty-seven per cent were males, and the median age was 53 (IQR = 37–62) yr. The haematological diagnoses included non-Hodgkin’s lymphoma (28%), acute myeloid leukaemia (27%), acute lymphoblastic leukaemia (11%), Hodgkin’s lymphoma (10%), and others (24%). Twenty-eight per cent of the patients had previously received haematopoietic stem cell transplantation (HSCT), 81% allogeneic and 19% autologous, and 36% patients had recent chemotherapy (within 4 weeks of ICU admission). Six patients also had human immunodeficiency virus disease.

Acute respiratory failure (45%), systemic sepsis (24%), neurological (10%), and cardiac illness (7%) accounted for acute decompensation and admission to ICU, with a median APACHE II score of 25 (IQR = 22–30) representing severe acuity of illness. Forty-two per cent were neutropenic (neutrophils ≤ 1 × 10\textsuperscript{9} litre\textsuperscript{−1}) at admission. Invasive mechanical ventilation was required in the majority of patients (73%), and renal support in 39%.

The presence of infection was identified in 69 out of the 97 patients (71%); 56 had confirmed infection and 13 were presumed to have an infection on clinical grounds. One, two, or three types of organism were identified in 32, 15, and 6 patients, respectively. Coagulase-negative Staphylococcus (n = 21), Pseudomonas aeruginosa (n = 15), and Enterococcus (n = 14) were the most frequent bacterial isolates, others being Escherichia coli (n = 6), Stenotrophomonas maltophilia (n = 6), Klebsiella sp. (n = 5), Acinetobacter sp. (n = 5), and Staphylococcus aureus (n = 3). Cytomegalovirus (CMV), Human Herpes Virus 6 (HHV6), and Epstein–Barr virus (EBV) were identified in 17, 5, and 5 patients, respectively, while fungal infection with Candida species were present in six patients.