

Spontaneous breathing in Ehlers-Danlos syndrome using Paedfusor model

Case Report



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Ehlers-Danlos syndromes are a heterogeneous group of heritable connective tissue disorders caused by mutation in genes collagen-related. Common features are: articular hypermobility, skin extensibility, and tissue fragility.

Patient was affected by EDS-HT, his mother also had this specific subtype of EDS. EDS-HT and joint hypermobility syndrome are clinically overlapping connective tissue disorders. These are featured by joint hypermobility, musculoskeletal pain, and minor skin alterations

A 4 year old boy affected by EDS-HT was scheduled for left orchidopexy. He showed kyphoscoliosis, thin hair, joint hypermobility and velvety skin. His mother was also affected by Ehlers-Danlos syndrome hypermobility type. Patient had all major and minor criteria to diagnose Ehlers-Danlos but there was no genetic alteration.

After inhalatory induction, IV access was secured, Propofol infusion using TCI model Paedfusor was started.

In order to obtain spontaneous ventilation with adequate hypnosis, concentration was set at 5 $\mu\text{g/mL}$ to achieve optimal depth of anesthesia, its concentration was titrated according to surgical procedure.

Ileo-hypogastric block was performed to obtain intraoperative and post-operative analgesia, using levobupivacaine 0.25% 0,5ml/kg.

Child maintained spontaneous ventilation and vital signs were stable throughout surgical procedure. Post-operative period was uneventful, no pain or discomfort was reported, he was discharged without complication.

EDS may be a challenging syndrome, we showed that intravenous anesthesia using Paedfusor mode and adding peripheral nerve block are safe and effective, moreover perioperative complications are less frequent because airway management is less invasive.



Fig.1 Child in spontaneous breathing with Paedfusor model, meanwhile peripheral nerve block is performed

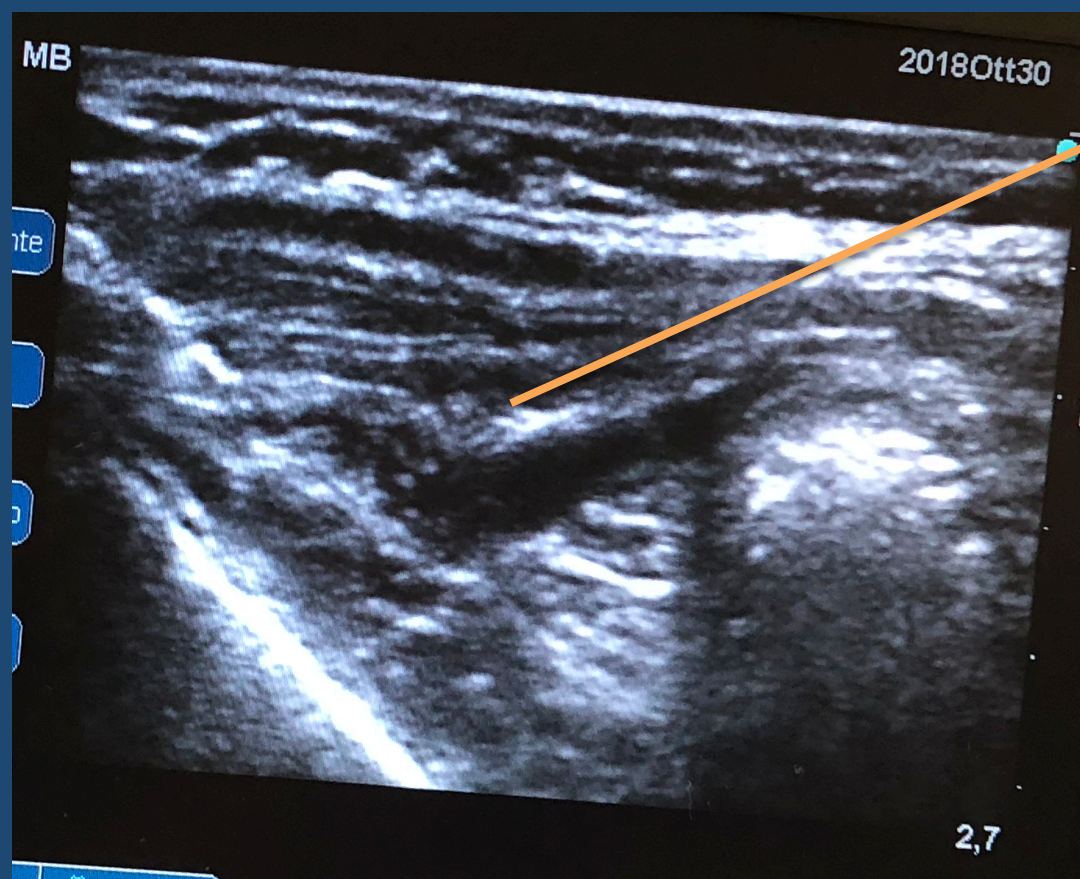


Fig.2 US-guided Ileo-hypogastric block