Ultrasound Guided Supraclavicular Block in Poland Syndrome: A Safe Alternative to General Anaesthesia

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Abstract: Poland syndrome is an uncommon, congenital anomaly characterized by multiple systems and organs involvement like musculoskeletal, lungs, heart, kidneys and haemopoietic system with absence of functional disability. Right sided thorax deformity is the most common feature of this syndrome with the absence of underlying bones and muscles making this condition similar to open chest along with ipsilateral hand anomalies such as brachydactyly, syndactyly or ectodactyly [1]. The incidence of this syndrome is 1 per 30,000-50,000 live births [2].

Patients with Poland syndrome require anesthesia during surgical procedures to correct the associated deformities or the organ systems associated with the disease process. Literature shows that these patients may be more susceptible for malignant hyperthermia due to associated musculoskeletal abnormalities under general anesthesia [3]. Moreover respiratory problems and hypoxia in the perioperative period may be related to associated paradoxical breathing, lung hypoplasia and chest wall abnormalities in otherwise asymptomatic patient at rest. We report a diagnosed case of Poland syndrome presented for syndactyly release.

Keywords: USG blocks, Supraclavicular approach.

CASE REPORT

A 7 years old 20 kg female presented for syndactyly repair of right hand. The birth history revealed that she was born full term with birth defect of right side thorax and ipsilateral hand so a diagnosis of Poland syndrome was at the time of birth. There was no history of previous surgery. Preoperative physical examination revealed no abnormality of any other system except chest wall and hand deformity of right side.

After thorough discussion with the parents about the risk and benefits of general anesthesia and written informed consent by the parents the Peripheral nerve block using ultrasound was planned.

Premedication with oral midazolam (15mg) and simultaneous application of EMLA cream on the dorsum of left hand was done and, patient was shifted to the operation theater after 30 minutes.

In the O.T all the routine monitors were applied and and a 22G peripheral intravenous line was secured without any difficulty. Injection Ketamin 1 µg kg-1 was administered as a bolus followed by Injection Dexmedetomidine 1 µg/kg over 10 minutes. Maintenance of anesthesia was done using Dexmedetomidine infusion 0.5µg/kg/hr and stopped 10 minutes before the completion of surgery. A supplemental oxygen via a nasal cannula was delivered at 2 litres / minutes.

Ultrasound guided identification of supraclavicular brachial plexus was performed using Linear array transducer (6-13 HZ) (Sonosite, Fujifilm , M Turbo). The probe was held in coronel–oblique plane in the supraclavicular fossa of the desired side and needle prick was performed using in-plane technique.

The brachial plexus bundle which appeared as hypoechoic nodules. 1 ml of 0.5% Injection ropivacaine was injected lateral to subclavian artery after negative aspiration followed by deposition of 15 ml 0.5 % Ropivacaine and spread of local anaesthetic was observed in realtime. The successful motor and sensory block of the upper limb was observed after 20 minutes of injection. The surgery and the post operative period was pain free and uneventful.

Our case report defines problems of general anesthesia in a child with Poland Syndrome. The primary concern in such cases are the problems of General anaesthesia with the use of succinylcholine and inhalational agents to trigger malignant hyperthermia [3]. Moreover under general anaesthesia with spontaneous ventilation the paradoxical respiratory movements would increase risk of inadequate ventilation often manifested as hypoxia requiring positive pressure ventilation even for short procedures [4]. So we planned supraclavicular brachial...
plexus block under sedation for the surgery avoiding general anaesthesia. Nerve stimulation-assisted peripheral nerve blocks (PNBs) is conventionally used technique, but is has high failure rate in the pediatric population. Although ultra sound guided supraclavicular brachial plexus block is ideal technique for shoulder, arm and hand surgeries in adults, it is not routinely used in paediatric patients due to risk of inadvertent vascular injury and pneumothorax [5].

More recently several studies have revealed U.S guided nerve blocks a safe technique even in children as its advantages includes direct visualization of anatomical structures and needle advancement thus decreasing complications and early functional recovery [6]. As we needed good sedation for our patient we decided to use dexmedetomidine and ketamine.

Existing literature reporting use of intravenous Dexmedetomidine and Ketamin combination for sedation are limited compared to the use of these drugs alone. Mester et al. used these drugs in combination for sedation successfully in paediatric patients undergoing cardiac catheterization [7].

Similarly Mc Vey et al. reported excellent procedural sedation using injection Dexmedetomidine and ketamine maintaining spontaneous respiration in paedric patients for lumbar puncture giving spinal anaesthesia [8]

Dexmedetomidine alpha -2 agonist, is FDA approved only for use in adult patients there are reports of successfully using it in paediatric age group for different clinical settings [9]. Although the use of
injection ketamine has advantage of rapid onset of sedation without bradycardia, side effects like emergence agitation, excessive salivation and sympathetic stimulation are prevented by the use of injection Dexmedetomidine along with conscious sedation and maintenance of spontaneous respiration. Our patient also received midazolam as premedication which might had further calmed our patient.

As there is always possibilities of hemodynamic instability and respiratory depression with these drugs we focused on standard monitoring, availability of resuscitation medications and equipments for airway management during procedural sedation.

CONCLUSION

As Poland Syndrome poses challenges to anaesthesiologist, our case report gives an insight that ultrasound guided supraclavicular block using procedural sedation can be used safely for these patients undergoing surgery of upper limb.

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