

Introduction

- Noonan syndrome is a rare genetic disorder involving pathologic changes to multiple organ systems, including chronic pain, posing a challenge to safe and effective anesthesia and analgesia for patients undergoing surgery
- Abnormal peripheral nerve anatomy including nerve fascicle and root hypertrophy has been described in patients with Noonan syndrome ^{1, 2}
- Little published describing the safety and efficacy of regional anesthetic techniques in this population
- We present a case of successful and safe post-operative regional anesthesia using a continuous femoral nerve catheter in a patient with Noonan syndrome found to have significant peripheral nerve hypertrophy on ultrasound

Written Health Insurance Portability and Accountability Act (HIPAA) authorization has been obtained from the patient for the presentation of this case report and ultrasound images.

Case Report

- 38-year-old female (52kg, 152 cm) with Noonan syndrome underwent scheduled left partial patellectomy from prior traumatic injury
- Received a single-shot femoral nerve block (22mL of 0.5% bupivacaine) performed under direct ultrasound guidance (Sonosite, L15-4) followed by an uncomplicated general endotracheal anesthetic
- Pre-procedural ultrasound of the femoral nerve found significantly hypertrophied nerve fascicles (Figure 1)
- Surgical procedure was well-tolerated without complications

- On POD#2, acute pain service placed a left femoral nerve catheter (17G Tuohy, 19G catheter) under direct ultrasound guidance with similar findings (Figure 2)
- A basal and patient-controlled regional anesthesia (PCRA) infusion of 0.125% bupivacaine was continued through POD #7 when the catheter was removed
- The patient reported significant subjective improvement in postoperative pain control and satisfaction with the addition of PCRA
- Daily OME was not significantly decreased
- There were no patient-reported adverse events related to the peripheral nerve catheter or PCRA infusion

Ultrasound Images

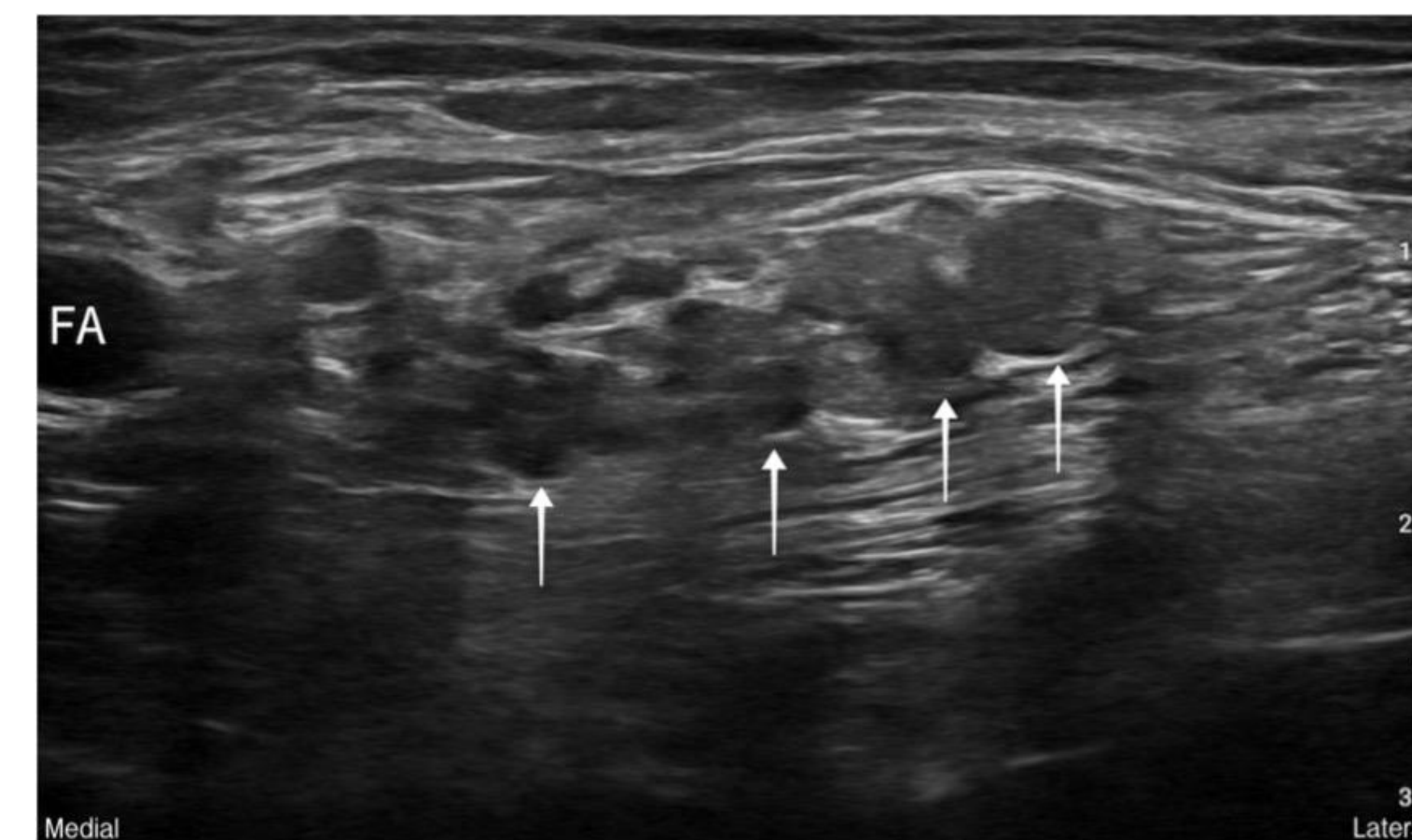


Figure 1
Transverse view of femoral nerve at the level of the inguinal crease and common femoral artery (FA) demonstrating significant nerve fascicle hypertrophy (arrows). The femoral nerve lies lateral to the common femoral artery.

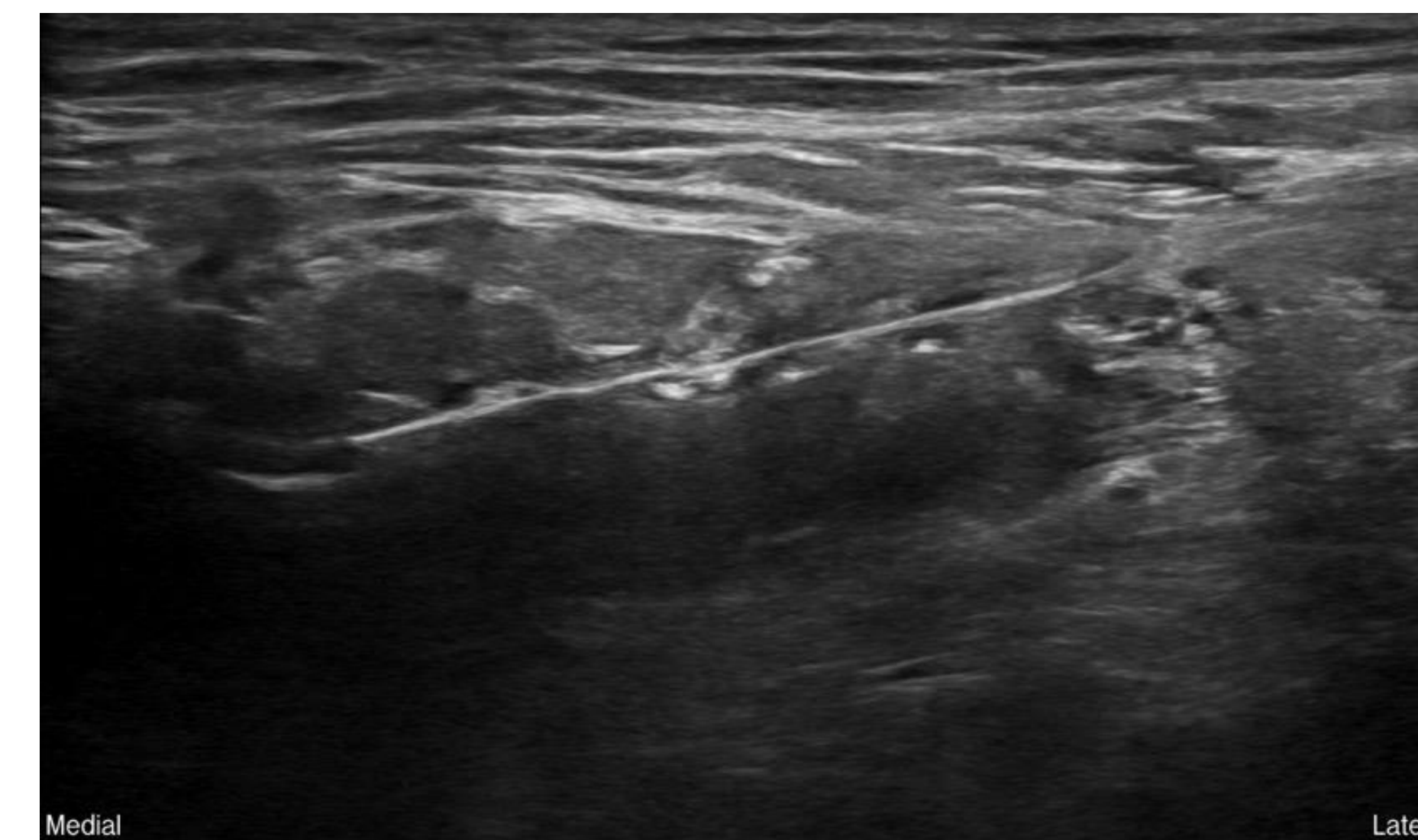


Figure 2
Transverse view of femoral nerve at the level of the inguinal crease demonstrating femoral nerve with block needle in position prior to placing a catheter underneath the nerve fascicles.

Conclusions

- Demonstrated safe and successful use of a continuous nerve catheter for patient with Noonan syndrome and abnormal peripheral nerve images
- Considerations included potential increased risk of nerve injury or intra-neural injection, local-anesthetic neurotoxicity, as well as poor absorption
- Similarities in peri-operative management for other polyneuropathies with peripheral nerve enlargement including Charcot-Marie Tooth and Neurofibromatosis type 1 ^{3, 4}
- Additional research is needed in a larger population, however regional anesthetic techniques should be considered on an individual basis

References

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