

# The Use of Peripheral Nerve Block and Intra-articular Steroid Injection for Pain Management in an Adolescent with Hutchinson-Gilford Progeria Syndrome: A Case Report

Marie Christelle S. Endencia, MD,<sup>1</sup> Karl Matthew C. Sy Su, MD<sup>1,2</sup> and Catherine Renee B. Reyes, MD<sup>1</sup>

<sup>1</sup>Department of Anesthesiology, College of Medicine and Philippine General Hospital, University of the Philippines Manila

<sup>2</sup>Department of Pharmacology and Toxicology, College of Medicine, University of the Philippines Manila

## ABSTRACT

Hutchinson-Gilford Progeria Syndrome (HGPS) is a rare genetic disorder causing accelerated aging and age-related pathologies. Weighing benefits and risks on doing surgical versus conservative pain management require multidisciplinary planning and consideration in HGPS patients. This presents a case of a 15-year-old patient with HGPS with severe pain from bilateral hip dislocation managed with peripheral nerve block and steroid injection. This afforded her immediate pain relief allowing her to undergo physical rehabilitation comfortably.

*Keywords: progeria, pediatric, anesthesia*

## INTRODUCTION

Hutchinson-Gilford progeria syndrome (HGPS) is a rare genetic disorder with a reported incidence of 1 in 8 million births.<sup>1</sup> Patients with progeria have characteristic features including alopecia, sclerodermatous skin, lipodystrophy, and osteolysis of clavicles, ribs, and phalanges.<sup>2,3</sup> They follow a normal psychomotor development but later develop multisystem comorbidities associated with aging such as coronary artery disease, cerebrovascular disease, hypertension, and atherosclerosis.<sup>2-4</sup> There have been reports on the anesthetic management in patients with HGPS, however, there is little to no literature on the use of pure locoregional anesthesia for pain management in these patients.

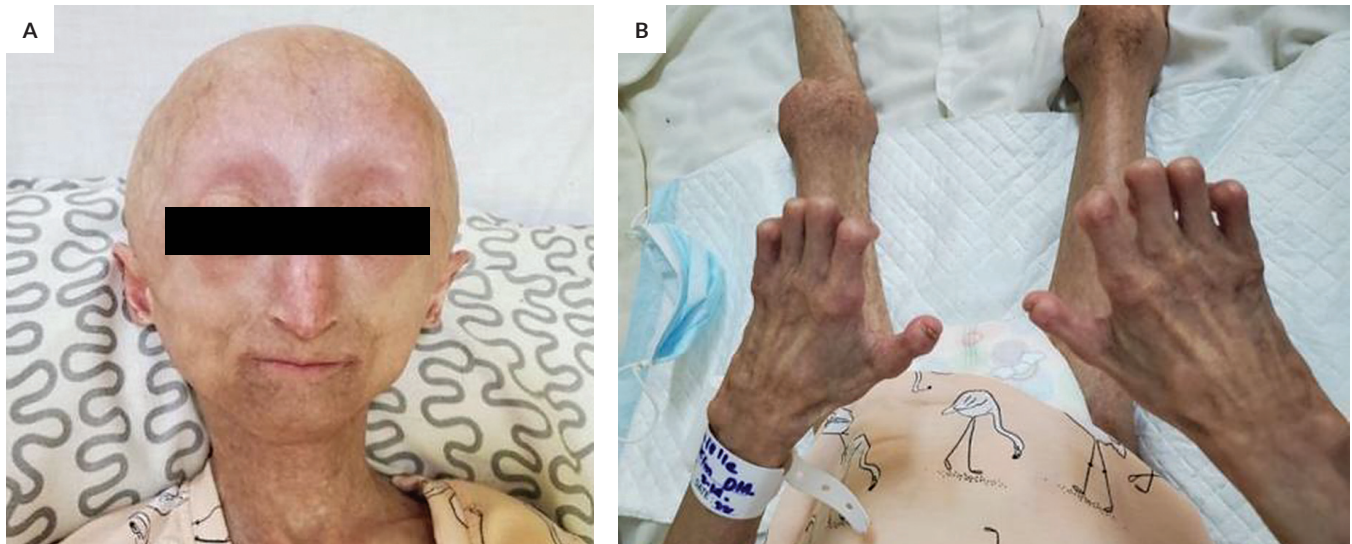
## CASE REPORT

A 15-year-old female with HGPS was referred for management of chronic bilateral hip dislocation. She was diagnosed with progeria at one year old through genetic testing after developing the characteristic facies of HGPS. One year before admission, she suffered a right hip dislocation after a fall which was managed with non-surgical reduction and has rendered her bedridden. In the same year, she underwent total hysterectomy and bilateral salpingo-oophorectomy under combined general and epidural anesthesia, during which, her left hip was spontaneously dislocated in surgery. She had an uneventful postoperative course but has been experiencing severe bilateral hip pain on minimal movement since her discharge, which prompted consult.



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Corresponding author: Marie Christelle S. Endencia, MD  
Department of Anesthesiology  
Philippine General Hospital  
University of the Philippines Manila  
Taft Avenue, Ermita, Manila 1000, Philippines  
Email: [msendencia1@up.edu.ph](mailto:msendencia1@up.edu.ph)  
ORCID: <https://orcid.org/0000-0003-2942-6534>



**Figure 1.** Characteristic phenotype of HGPS (alopecia, sclerodermatous skin, small face with pinched nose, decreased subcutaneous tissue, nail dystrophy).

The patient has a normal birth and developmental history with no other known systemic disease. She was started on Lonafarnib capsules at 3 years of age as maintenance medication.

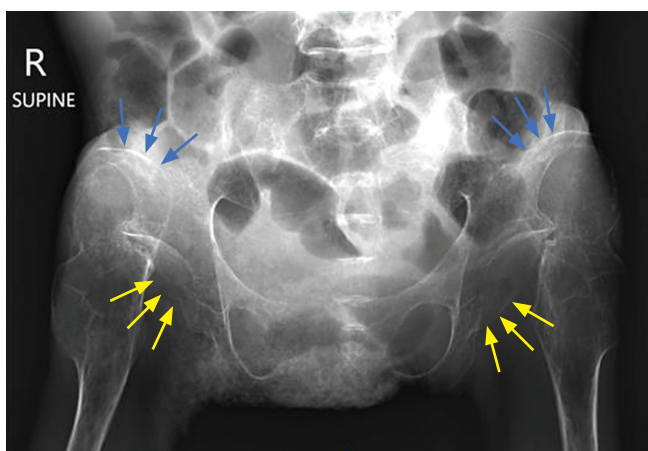
The long-term goal is to have the patient sit without pain to achieve community mobility with a motorized seating device. Surgery was the initial plan for the patient, however, due to her high-risk nature, a more conservative approach of doing a peripheral nerve block with intra-articular steroid injection on both hips was planned to enable her to do physical rehabilitation exercises comfortably.

The patient was admitted for elective peripheral nerve block and initiation of bedside physical rehabilitation. On examination, the patient was seen awake, conversant, on low back rest, with normal vital signs for age. She weighed

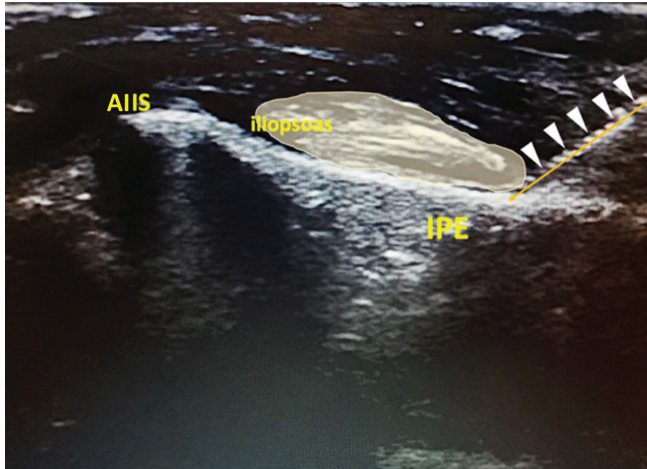
10.4 kg with typical progeria manifestations (alopecia, sclerodermatous skin, small face with pinched nose, crowded teeth, decreased subcutaneous tissue, nail dystrophy, and narrow upper thorax) (Figure 1). Other systemic physical exam findings were unremarkable, but she had severe pain (NPS 7-9/10) on both hips even on minimal movement.

On work-up, laboratory blood tests were unremarkable. Electrocardiogram revealed left ventricular hypertrophy with nonspecific ST-T wave changes; chest radiography showed cardiomegaly; 2D echo and doppler studies showed left ventricular hypertrophy with normal cardiac functions; and handheld spirometry revealed mild obstructive lung disease. On hip radiography, she had superior dislocation of both femoro-acetabular joints with sclerotic changes (Figure 2). She was then scheduled for nerve block the following day.

On the day of the procedure, the patient was calm and cooperative. Standard ASA monitors were applied, and she was sedated using fentanyl 0.5 mcg/kg intermittent boluses as needed. Bilateral hip pain on movement excluded the use of blocks that require repositioning; therefore, a single-shot pericapsular nerve group (PENG) block was planned for both hips with supplemental intra-articular triamcinolone acetonide injection. Observing aseptic technique, PENG block was done by positioning a high-frequency linear probe parallel to the inguinal crease at the level of the anterior superior iliac spine. The transducer was gradually moved medially along this axis until the anterior inferior iliac spine (AIIS), iliopubic eminence (IPE), and psoas tendon were identified. After anatomic landmarks were identified, local anesthesia infiltration on skin was given using lidocaine 2%, then a 50 mm 22-gauge needle was introduced using ultrasound-guided in-plane technique (Figure 3). After negative aspiration, 7 ml of a 0.15% bupivacaine solution with 4 mg dexamethasone was administered on both hips



**Figure 2.** Hip x-ray showing bilaterally dislocated femoral heads (yellow arrows: acetabula; blue arrows: femoral heads).



**Figure 3.** Sono-anatomy of PENG Block (white arrow heads showing direction of the needle).

AIIS - anterior inferior iliac spine; IPE - iliopectineal eminence

with noted spread of local anesthesia between the IPE and psoas tendon on ultrasound. Additional intra-articular triamcinolone acetonide was injected on the left hip to augment analgesia but was not done on the right hip due to diminished femoro-acetabular joint. The patient was fully awake after the procedure and was sent back to the ward to recover.

After the procedure, there was a significant improvement in bilateral hip pain from NRS 7-9/10 to NRS 0-2/10 on passive movement. The patient was able to do passive range of motion exercises without any pain and was able to move herself to different sitting heights comfortably. The patient reported no pain (NRS 0-1/10) throughout her physical therapy exercises and was able to do assisted active movements comfortably throughout her hospital stay. The patient was discharged after one week.

## DISCUSSION

Hutchinson-Gilford Progeria Syndrome (HGPS) is a rare genetic disorder resulting in accelerated aging, severe growth failure, and early-onset atherosclerosis caused by a mutation in the LMNA gene.<sup>1,5</sup> Handling patients with HGPS for possible surgery require careful and often, multidisciplinary planning to anticipate possible difficulties that may be encountered in the perioperative course.

The patient was initially referred to Anesthesia service for surgical correction of her bilateral dislocated hips. However, multiple anesthetic concerns were considered before her operation. The complications associated with the advanced physiologic age in HGPS patients are one of the foremost concerns. They gradually develop elevated blood pressure starting 6 to 8 years of age<sup>3</sup> and later have other cardiovascular abnormalities such as valvular heart disease or left ventricular remodeling during their second

decade of life<sup>6</sup>. They should therefore undergo preoperative cardiac investigations<sup>7</sup> and be referred to pediatric and adult cardiology specialists to assess cardiac risk.

Pulmonary function must also be adequately assessed in HGPS. The shortened telomere length<sup>8</sup> is similar to the geriatric population and may be associated with decreased lung function and increased risk of obstructive pulmonary disease<sup>9</sup>. Protective ventilatory techniques in patients with obstructive lung disease<sup>10,11</sup> should be considered if they will be mechanically ventilated intraoperatively.

A salient consideration in anesthetizing these patients is the possibility of a difficult airway. Typical airway features in progeria including retro and/or micrognathia, mandibular hypoplasia, small mouth and glottic opening, and high arched palate<sup>5</sup> are significant concerns when planning to secure their airway. Preparation of advanced airway equipment and alternative techniques to secure the airway must be ensured prior to induction.

Other considerations in progeria are transfers and positioning which should be done with care due to susceptibility to injuries and skin avulsions due to sclerodermatous skin.<sup>12</sup> They are also prone to hypothermia due to decreased subcutaneous fat and alopecia<sup>13</sup>, and hypoglycemia due to increased insulin levels<sup>14</sup>.

The benefits and risks of surgery were weighed against the goal for this patient. Since community ambulation through a seating device was the long-term plan, the treatment was aimed toward the control of bilateral hip pain to facilitate physical therapy and allow her to sit and transfer with ease. The plan was then shifted to a more conservative approach to pain relief through peripheral nerve block and intra-articular steroid injection.

The pericapsular nerve group (PENG) block is a new technique used in patients with hip fractures. This fascial plane block involves the deposition of anesthetic in the fascial plane between the superior pubic ramus and iliopsoas muscle to block the articular branches supplied by the femoral (FN), obturator (ON), and accessory obturator nerves (AON).<sup>15</sup> In contrast to the femoral nerve block and fascia iliaca compartment block, the PENG block provides more robust analgesia by blocking the ON and AON which innervates the anterior hip capsule with motor-sparing effects.<sup>16</sup>

Case reports on the use of PENG block in pediatrics all showed satisfactory hip analgesia with preserved motor function.<sup>16-18</sup> The motor-sparing effect of the block is beneficial because the patient will be able to use her quadriceps during assisted active movements during physical therapy. A disadvantage of a single-shot peripheral nerve block is its limited analgesic duration. Although some studies have concluded the safety of continuous peripheral nerve catheters<sup>19,20</sup>, they still reported a 12.1-14.4% complication rate<sup>19,20</sup> including accidental catheter dislodgement or removal, infection, catheter leak, and local anesthetic toxicity. One case report documented success in the use of continuous PENG block for perioperative analgesia in a child<sup>21</sup>, but the



applicability of their continuous technique may not be valid in this patient who required bilateral hip analgesia. Because the patient will need an extended duration of analgesia for her physical therapy, the duration of the single-shot block was prolonged by adding dexamethasone to the local anesthetic. Despite limited recommendations on the use of adjuncts in peripheral nerve block in pediatrics<sup>22</sup>, meta-analyses have shown that perineural dexamethasone safely lengthens the duration of analgesia of peripheral nerve blocks<sup>23,24</sup>, and therefore fulfills the fundamental requirements on the use of adjuvant drugs in pediatric regional anesthesia<sup>22</sup>.

In addition to the PENG block, an intra-articular steroid injection was also performed to give longer hip analgesia. Intra-articular steroid injection is a minimally invasive pain management modality used in different musculoskeletal conditions.<sup>25</sup> It has been used in pediatric and adult cerebral palsy patients with hip dislocations demonstrating an effective decrease in pain scores<sup>25,26</sup> and was the treatment of choice in patients with shorter life expectancy or when there were general anesthesia contraindications<sup>26</sup>. One disadvantage of this technique, however, is the longer onset of action, generally within 24–48 hours from injection<sup>24–26</sup> but analgesia may last up to 3–5 months<sup>26</sup>. The combination of a regional block and an intra-articular injection was a reasonable treatment plan for this patient to provide immediate analgesia to initiate physical rehabilitation and afford longer-lasting pain relief to allow community ambulation.

## CONCLUSION

Hutchinson-Gilford Progeria Syndrome (HGPS) is a genetic disorder with a distinct phenotype that is challenging to anesthesiologists. We presented an adolescent with HGPS who consulted for management of bilateral hip pain from hip dislocation. After weighing the risks and benefits, bilateral pericapsular nerve group (PENG) block and intra-articular steroid injection were planned to afford pain-free sitting and ambulation through a seating device. This provided immediate and long-lasting analgesia that allowed physical therapy initiation until her discharge.

## Informed Consent

Written consent and assent from parent and patient, respectively, have been obtained to post photographs for educational purposes.

## Statement of Authorship

All authors certified fulfillment of ICMJE authorship criteria.

## Author Disclosure

All authors declared no conflicts of interest.

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