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The Anesthetic Challenges of Caring for a Pediatric Patient With Incontinentia Pigmenti: A Case Report

Shabaaz M. Baig, BA, and Shridevi Pandya Shah, MD

Incontinentia pigmenti (IP) is a rare X-linked dominant disorder. We present a case of an infant with IP who was brought to the operating room for panretinal diode photocoagulation under general anesthesia. The anesthesia team was unable to obtain intravenous access even with instruments such as a vein finder and ultrasound. Anesthesia for IP patients also poses challenges such as prevention of the oculocardiac reflex, obesity and airway management, and preemptive measures for intravenous access due to skin manifestations. Patients with IP may present with many challenges for the anesthesiologist during all phases of anesthetic management. (A&A Practice. 2021;15:e01384.)

GLOSSARY

EQUATOR = Enhancing the Quality and Transparency of Health Research; **GERD** = gastroesophageal reflux disease; **IP** = incontinentia pigmenti; **OCR** = oculocardiac reflex; **OR** = operating room

Incontinentia pigmenti (IP) is an X-linked dominant disorder with primarily cutaneous manifestations but also wide-ranging neurological, ocular, and dental issues.[1–5] The disease is caused by a defect in the IKBKG gene on the X chromosome.[1,2] Cutaneous aspects of the disease involve mutated and normal cells competing in the skin along the Blaschko lines.[1,2] These skin findings are the sole major diagnostic criteria for IP and can be graded into 4 stages (Table).[1,2] The lesions go through these stages as the patient grows older. Other criteria for diagnosis include ocular, neurological, and dental anomalies.[2,6] Common abnormalities reported in the literature include strokes, seizures, cataracts, and macular vasculopathy.[1–3,7] Although cardiac complications are rare, pulmonary hypertension may occur.[1] Ocular anomalies, though not a concern in patients with mild IP, may require intervention to prevent loss of vision.[2,6] Similarly, neurological abnormalities may be significant in IP due to the potential of irreversible damage.[1] Some patients with mild disease may go through life without realizing they have the disease.

These disease manifestations are important to consider for anesthetic management in patients with IP. In this report, we highlight several anesthetic challenges for an infant with stage 2 IP. This article adheres to the applicable Enhancing the Quality and Transparency of Health Research (EQUATOR) guidelines. Written Health Insurance Portability and Accountability Act authorization was obtained from the patient's family.

From the Department of Anesthesiology, Rutgers New Jersey Medical School, Newark, New Jersey.

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Address correspondence to Shridevi Pandya Shah, MD, Department of Anesthesiology, Rutgers New Jersey Medical School, 185 S Orange Ave, MSB-E 577, Newark, NJ 07103. Address e-mail to pandyas1@njms.rutgers.edu.

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CASE DESCRIPTION

Our patient was a 7-month-old girl born at 37 weeks via cesarean delivery with an uneventful gestational history. The patient was diagnosed with IP, adrenal insufficiency, gastroesophageal reflux disease (GERD) requiring nasogastric tube feeding, multiple strokes, bilateral cataracts, and neovascularization. Her medications included hydrocortisone 1.5 mg orally daily, levetiracetam 150 mg twice daily, and ranitidine 7.5 mg twice daily. She was brought to the operating room (OR) for a complete eye examination and panretinal diode photocoagulation under general anesthesia.[8]

The patient was first evaluated in the preadmission testing clinic. She was found to have lesions consistent with the second stage of IP (Figure) and neurological and ocular abnormalities. A few puncture marks were seen on her extremities secondary to attempts to gain intravenous access during magnetic resonance imaging about a week ago. The patient displayed a typical Cushingoid appearance, with moon facies and a buffalo hump in addition to edematous, flaky, hyperpigmented lesions on all extremities. The patient's weight was 8.4 kg. We suspected that intravenous access would be challenging and arranged to have a vein finder and ultrasound in the OR.

The patient was brought to the OR and a forced air warming blanket was placed underneath her. In the OR, standard American Society of Anesthesiologists monitors were placed. Because the patient's GERD was well controlled, we proceeded with a slow inhalation induction and hoped to be able to visualize veins after induction-associated vasodilation. Air exchange was maintained without an oral airway, and we planned to maintain spontaneous breathing during mask ventilation to avoid possible abdominal insufflation that can occur with positive pressure ventilation. The patient received intramuscular atropine because anesthetic depth was established to avert bradycardia resulting from sevoflurane use and to prevent the oculocardiac reflex (OCR).

Despite use of vein finder and ultrasound, we were unable to establish intravenous access, even after repeated attempts by multiple pediatric anesthesiology personnel. The patient's severe Cushingoid features made exposure of

Table. Skin Presentations of Incontinentia Pigmenti	
Stage 1	Seen at birth or within first 2 wk with patterned vesicles and/or pustules usually overlying an erythematous base and developing along the lines of Blaschko
Stage 2	Seen between 2 and 8 wk of life when the lesions become more papular or crusted
Stage 3	The hyperpigmented stage between 6 and 12 mo, noted with brown or gray-brown macules in a linear and/or swirling pattern along the lines of Blaschko. Typically lasts through early adolescence or can persist into adulthood
Stage 4	Hypopigmented and slightly atrophic linear macules and patches. Do not occur in the majority of patients

her neck difficult. Groin access was not considered due to the increased risk of infection, and the peripheral and brief nature of her scheduled procedure. Furthermore, we opted against central line placement because we believed that the potential risks of the procedure outweighed the benefits. An interosseous line was available in the OR in case of emergency.

Although postponing the case until her skin lesions and overall condition had improved was discussed with the management team, the surgeon wished to proceed, and the anesthesiology team agreed that it was appropriate. Subsequently, complete eye examination and laser photocoagulation were performed without complications. Though such procedures are typically not painful, inhalational anesthesia was supplemented with a nasal fentanyl dose of 0.1 µg/kg and 30 mg/kg of rectal acetaminophen. The patient was able to resume oral intake soon after the procedure, and recovery was uneventful.

Two weeks later, the patient returned to the OR for cataract extraction and vitrectomy. Since she was still displaying persistent hyperpigmented skin lesions and blisters over her extremities, she was admitted to the hospital the night before surgery to obtain intravenous access in advance. Although 1 peripheral intravenous access was established, a right internal jugular central line was placed in the preoperative period by the interventional radiologist. The decision to obtain this additional access was made in conjunction with the ophthalmic team to ensure optimal hydration and pain control and to continue medication without interruption.

DISCUSSION

Even though there are publications describing IP and its pathologies, there is no written work discussing the anesthetic management of severe cases of patients with IP who need timely intervention. Patients with cutaneous manifestations who are on chronic steroids may pose additional challenges, as our case demonstrated.

Patients with IP benefit from a visit to the preadmission testing facility due to potential multisystem involvement. Our patient's history warranted some unique considerations during the perioperative period. One of the most significant problems faced by our team was difficulty in obtaining adequate intravenous access. When examining patients with IP, anesthesiologists should carefully assess intravenous access points for patency. If patency cannot be confirmed, other measures including use of a vein finder, ultrasound guidance, or central line placement should be considered.

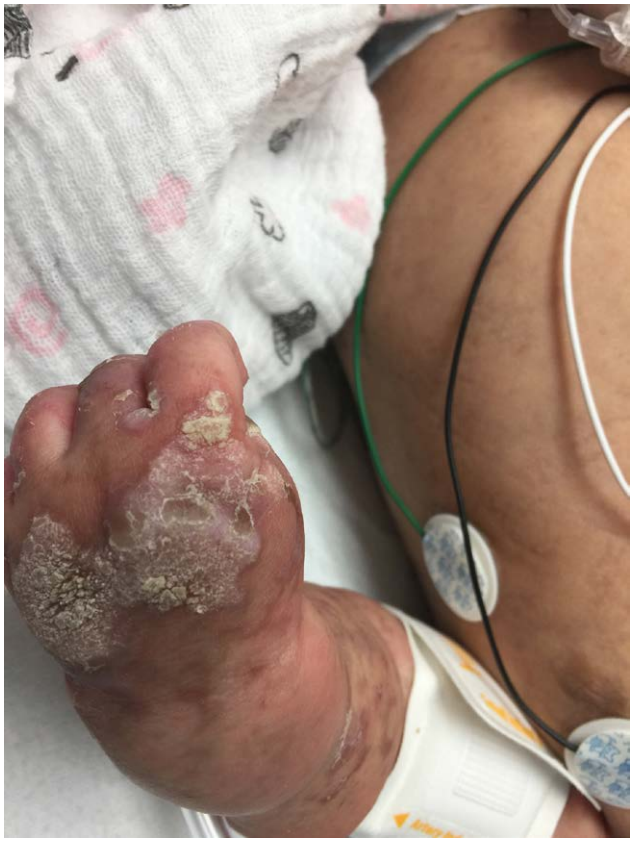

Figure. Our patient demonstrated characteristic stage 2 skin lesions of incontinentia pigmenti. Papular, crusted lesions were visible on all the patient's extremities.

As seen in our patient, long-term usage of steroids can lead to significant skin changes, which, when combined with preexisting IP lesions, can further increase the difficulty of gaining intravenous access. Steroid use may also lead to classic Cushingoid signs, such as obesity, buffalo hump, moon facies, and perioperative hemodynamic instability. Given our patient's history of adrenal insufficiency and daily steroid treatment, additional steroids were administered preoperatively.

According to Patak et al,[9] obesity is associated with significantly more difficulty in establishing intravenous access in children. This study, as well as research done by Petroski et al[10] reported that difficult intravenous access was most common at very young ages,[9] especially in the 7- to 11-month-old group.[9] Although groin access was not attempted in this patient, ultrasound examination of the neck and groin veins should be performed to determine vessel patency. Furthermore, these patients should also be thoroughly examined for cardiovascular abnormalities. Since pulmonary hypertension is a rare complication of IP,[1] an echocardiogram should be considered in symptomatic patients.

Perioperatively, active measures must be taken to keep the patient warm. Forced air warming and increasing room temperature should be adequate measures to achieve that goal. Depending on the IP skin lesions stage, it can be challenging to place a blood pressure cuff or electrocardiographic monitoring leads, and indeed it was difficult in our patient due to her widespread cutaneous manifestations. These patients

should be positioned in such a way that affected areas are protected as much as possible to avoid skin damage. An additional perioperative consideration with obesity is airway management. Our patient was not obese but her obvious Cushingoid features were alarming. While our patient exhibited no difficulties with mask ventilation, the ease of airway access must be monitored carefully.

A more specific potential complication during these cases can be the OCR, which is triggered by direct traction or pressure on the eye leading to bradycardia. If the OCR does occur, removal of traction is the initial effective measure. Intravenous atropine can also be administered to dampen the effects of the OCR.[11] In addition, injection of local anesthetic into the extraocular muscles can avoid further occurrences. Another preventive measure is a retrobulbar block.[11] This option was not a safe option in our case due to the lack of intravenous access. Furthermore, we determined that there was a lower likelihood of activating the OCR in this patient due to lack of direct traction on the extraocular muscles. Thus, both the surgeon and anesthesiologist determined it was reasonable to proceed with the case without intravenous access. Patients with IP may have postoperative concerns, including adrenal insufficiency, failure to thrive secondary to GERD, and difficult to manage pain. If patients are on steroids preoperatively, their usual steroid regimens are continued postoperatively.

IP requires specific preoperative and perioperative considerations. This disease has several important anesthetic implications including skin manifestations preventing intravenous access, chronic steroid use and its effects, cardiovascular issues like pulmonary hypertension. Although we were able to manage this patient without intravenous access due to the relatively noninvasive and superficial nature of her surgery, this may not always be the case when more invasive procedures are planned. Therefore, vigilance is required to avoid potential perioperative complications and procedural cancellations in patients with IP.

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Contribution: This author helped with writing the manuscript, literature review, and reviewing and revising the final manuscript.
Name: Shridevi Pandya Shah, MD.
Contribution: This author helped with writing the manuscript, providing initial editing and analyses, and reviewing and revising the final manuscript. This author was also the attending pediatric anesthesiologist for this case.
This manuscript was handled by: BobbieJean Sweitzer, MD, FACP.

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