Since its description by Behan in 2003, the keystone flap has gained wide popularity as a reliable and versatile technique for closing large soft-tissue defects in various regions of the body. It is named for its wedge shape, which resembles that of the critically important architectural stone found at the apex of Roman archways.^{1} Classically described in 4 variations, the keystone flap is a fasciocutaneous perforator island flap that is designed adjacent to the long axis of an elliptical defect. The variations are as follows: Type I—direct closure of primary and secondary defects without division of deep fascia, Type II—excess skin tension requires division of deep fascia along the trailing edge of the flap, Type III—2 keystone flaps, with one on each side of the primary defect, and Type IV—significant keystone flap undermining allowing for additional rotation or advancement.^{1} Dissection or direct visualization of perforating vessels is not required. As the flap is advanced, the resulting donor site is closed in a V-Y fashion at the trailing corners of the flap. This effectively narrows the donor site defect, enabling greater flap advancement and more even distribution of tension.^{1}

The keystone design perforator island flap has been reported to minimize the requirements for skin grafting and to be less technically demanding than other locoregional perforator flaps or microvascular free tissue transfer options, while maintaining favorable reconstructive outcomes.^{2} We present a case in which a type IV keystone design perforator island flap was used for closure of a posterior trunk defect following resection of a massive venous malformation in an 8-week-old infant with blue rubber bleb nevus syndrome (BRBNS).

**CASE REPORT**

The patient was a full-term male neonate (41 weeks, 3003 g) born via spontaneous vaginal delivery who was found to have a large, exophytic mass in the lower thoracic region, which occupied approximately one-third the surface area of the posterior trunk. Several small, bluish, papular skin lesions were also observed to be scattered sporadically across the body, involving the palms and soles (Fig. 1). MRI on day 2 of life demonstrated a large venous malformation involving the subcutaneous tissues of the back from T7 to T12. The malformation extended deeply through the paraspinal musculature to involve neural foramina on the right and continued into the posterior mediastinum. This neonatal patient was referred to our institution for evaluation by our interdisciplinary Vascular Anomalies Team, where he was subsequently diagnosed with BRBNS and started on sirolimus to minimize the risk.
of bleeding from gastrointestinal blebs. As a consequence of extensive phleboliths within the large venous malformation, the patient was found to have a severe consumptive coagulopathy requiring regular transfusions of fresh frozen plasma and cryoprecipitate until surgery could be arranged.

At 8 weeks of age, the patient underwent surgical excision of the exophytic portion of the large posterior trunk venous malformation for definitive treatment of his consumptive coagulopathy. This resulted in an approximately 8 cm by 10 cm defect of skin and subcutaneous tissue, with the transected stalk of the venous malformation exposed centrally at the base of the wound (Fig. 2). Immediate reconstruction commenced with a left paraspinous muscle flap to obtain total muscular coverage of the transected stalk of the venous malformation. A left latissimus dorsi myocutaneous flap was then widely elevated to maximize advancement of the medial edge of the primary defect. Superior and inferior medial corners of the defect were then temporarily stapled together, resulting in standing cutaneous deformities and an overall decrease in size of the residual defect to approximately 5 cm by 5 cm. A keystone design perforator island flap from the right flank was planned. Three perforators were identified using a handheld Doppler, and a 5 cm by 9 cm keystone flap was designed adjacent to the defect to include these perforators (Fig. 3). Dissection was carried out down to the level of the deep fascia of the chest wall. The deep fascia was then incised circumferentially, and the flap was partially undermined in the subfascial plane medially to maximize medial advancement. After wide undermining of the donor site wound edges, the superior and inferior corners of the donor site wound were closed in a V-Y fashion. The keystone flap was then advanced into the defect and inset over closed suction drains. Lastly, standing cutaneous deformities were excised superiorly and inferiorly from the medial edge of the primary defect, giving the appearance of additional Y-shaped closures (Fig. 4).

The patient required one unit of fresh frozen plasma transfusion and vitamin K supplementation in the initial postoperative period, and the consumptive coagulopathy completely resolved by postoperative day 5 without further treatment. The patient was restricted to prone or left lateral decubitus positioning for the first postoperative...
week to avoid direct pressure on the keystone flap, which healed without incident (Fig. 5).

DISCUSSION

BRBNS is a rare syndrome characterized by multiple venous malformations that can involve any tissue. The skin (78%) and gastrointestinal tract (89%) are most commonly affected. While bleeding from gastrointestinal lesions can be problematic later in life, this patient presented with a life-threatening consumptive coagulopathy due to pooling of blood in the large posterior trunk venous malformation.

An interdisciplinary team approach has become the standard of care in managing patients with complex vascular anomalies. In this case, our Vascular Anomalies Team was critical in mitigating the risk of hemorrhage, both medically and surgically. Medical management with sirolimus was initiated at the time of diagnosis to limit the frequency and severity of future gastrointestinal bleeds, which are almost ubiquitous in patients with BRBNS. Sirolimus interferes with the function of mammalian target of rapamycin, inhibiting the mammalian target of rapamycin–mediated signal-transduction pathways, thereby resulting in the arrest of cell cycle in G1 phase in various cell types. Aggressive surgical debulking of the large posterior trunk venous malformation was required to definitively treat the patient’s consumptive coagulopathy. The plastic surgeon’s integral role on this team was to provide an aesthetically acceptable reconstruction of the large posterior trunk defect that resulted from the resection.

Keystone flaps in infants have been previously reported in the context of myelomeningocele defects. To our knowledge, this is the first report describing utilization of a keystone flap for reconstruction of a posterior trunk defect after resection of a large venous malformation in an infant. Several other approaches to posterior trunk reconstruction have been described, including various rotation, advancement or transposition flaps, bilobed flaps, Z-plasties, dorsal intercostal artery perforator propeller flaps, and reverse turnover latissimus dorsi muscle flaps. A keystone design perforator island flap was chosen in this case due to its reliability and relative ease of execution. Perhaps the greatest advantage of the keystone flap is the robust vascular supply, which need not be identified or dissected during flap elevation. This translates to decreased surgical complexity and shorter operative times. The long-term reconstructive outcome for this patient was found to be satisfactory.

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