Case 1: Neonatal labial swelling

A three-week-old girl presented to the emergency department with a swollen purple labia (Figure 1) that had developed over a 24 h period. She was feeding and voiding well, and there was no recent change in formula or brand of diapers. There was no history of trauma. The baby was somewhat irritable, and her diaper area was very tender to touch. She was afebrile, and had no sick contacts. She had no cyanosis or respiratory distress. Her medical history showed that she was born to a 19-year-old gravida 2, para 2 mother who had limited antenatal care. The baby was born by planned caesarian section due to active herpes lesions. There was no rupture of membranes at the time of delivery. Neither mom nor baby received antiviral medications. The baby had a heart murmur detected at birth, and was diagnosed with double-outlet right ventricle. Surgery was scheduled for a later date. She was not receiving any medications and had no known allergies. The Children's Aid Society had previously been involved with the family, and the family physician described the social circumstances as extremely high risk. The child was referred to the on-call paediatrician at the regional sexual assault and child abuse centre because of concerns of nonaccidental injury.

On initial physical examination, her heart rate was 140 beats/min, respiratory rate was 32 breaths/min, temperature was 37.7°C, oxygen saturation was 99% and her weight was 3.042 kg. The patient's respiratory examination was normal. On cardiac examination, she had a pansystolic murmur, but normal cap refill and good pulses. She had no peripheral edema. Her abdomen was soft and nontender, with no hepatosplenomegaly. Her neurological examination was normal. Her skin examination was normal, apart from an impressive purple discolouration in the diaper area, with tender swollen labia (Figure 1). A blood sample revealed a white blood cell count of 12.8×10^9/L, with a 28% left shift; a hemoglobin level of 123 g/L and a platelet count of 391×10^9/L. The infant was admitted for further observation, and her clinical progression overnight led to the diagnosis.

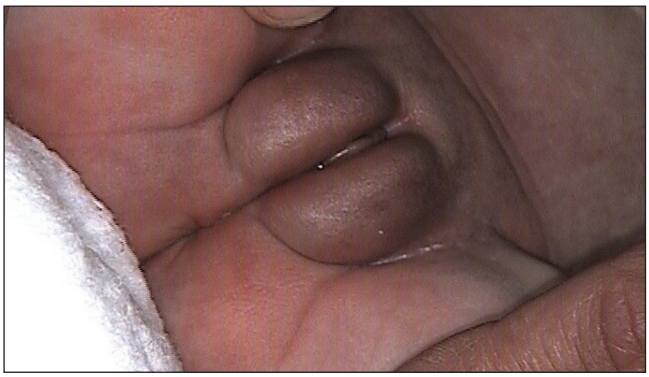

Figure 1) *Three-week-old girl with swollen purple labia*

Case 2: Unilateral facial flushing with eating

A previously healthy three-year-old girl presented with a one-week history of an episodic erythematous patch extending from the corner of the mouth to the right cheek. The eruption occurred following eating, but on occasion would occur with teeth brushing and finger sucking. There was no associated pruritus, scale, vesicles, pustules, burning or scarring. The eruption was not associated with sun exposure.

Although the eruption seemed to be primarily associated with eating, it was not linked to any particular food. The flushing lasted from 30 min to 60 min, and did not show a response to either a midpotency topical corticosteroid or a topical antibiotic.

On review of systems, there were no associated symptoms of facial swelling, wheezing, urticaria, vomiting or abdominal pain. There was no history of involvement in other sun-exposed areas. The patient's past medical history is only significant for a ventricular septal defect, previously spontaneously closed. Of note, there was no history of traumatic delivery.

The patient's physical examination revealed a well-appearing young girl. Her vital signs were normal. She had a blanching erythematous patch extending from the right cheek to the right corner of her mouth (Figure 1). The rest of her examination was normal.

Reviewing the patient's history and physical examination revealed the diagnosis.

Figure 1) *Patient's face at presentation demonstrating erythematous flushing over the right cheek*

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TABLE 1
Rochester criteria for febrile infants at low risk of having a serious bacterial infection

Age group	≤60 days
Medical history	Born at term age
	Home with or before mother
	No subsequent hospitalizations
	No perinatal, postnatal or current antibiotics
	No treatment for unexplained hyperbilirubinemia
	No chronic disease
Physical examination	Appears generally well with no evidence of skin, soft tissue, bone, joint or ear infection
Laboratory	
Total WBC count	5.0×10^9/L to 15.0×10^9/L
Bands	$<1.5\times10^9$/L
Urine	<10 WBC/HPF*
Stool (if diarrhea)	<5 WBC/HPF

**Refers to examination of centrifuged urine sediment, examined under 40× microscopy. HPF High-power field; WBC White blood cell. Adapted from reference 1*

CASE 1 DIAGNOSIS: SEPTIC SHOCK

Differential diagnosis at the time of admission included infection, cardiogenic edema and nonaccidental injury. Given that the child looked well, her rash did not look typical for infection and her bloodwork was remarkable only for the left shift, she was admitted for observation and no active treatment was started. However, 10 h after admission, the on-call paediatrician was paged urgently due to significant deterioration in the infant's respiratory status. On examination, the infant was tachypneic, cool, mottled and reacting poorly. The purple discolouration had turned to red, and migrated laterally and superiorly to the level of the umbilicus. A chest x-ray demonstrated cardiomegaly, but no abnormalities in her lung fields. She was intubated to stabilize her respiratory state. She had profound metabolic acidosis with a pH of 6.9 and a bicarbonate concentration of 3 mmol/L. Her mean blood pressure fell to 29 mmHg, and normal saline and vasopressors were administered. She was catheterized for urine, but had insufficient output to send a culture. She was not stable enough for lumbar puncture. Based on her clinical presentation, she was diagnosed with septic shock and treated with meningitic doses of ampicillin, cefotaxime and acyclovir. She required significant fluid resuscitation as well as inotropic support, but with aggressive treatment, she recovered fully from her illness. No organism was cultured.

The child presented with an unusual rash (Figure 1), but otherwise unremarkable physical examination, and quickly deteriorated into septic shock. With the unusual skin rash, and a high band count, the patient did not meet low-risk criteria for serious bacterial infection, according to the Rochester criteria (Table 1). However, a MEDLINE search of the literature (1950 to 2008) failed to reveal any case reports of purple labial discolouration with sepsis in infants. To avoid overdiagnosis of sexual abuse, several sources suggest a full and complete evaluation of perianal rashes. The vast majority of children referred for evaluation due to disclosure of sexual abuse have normal or nonspecific findings. Perianal streptococcal infection is common in infants and children. However, this typically presents as a bright red confluent rash, with or without impetiginous lesions. Candidal diaper dermatitis may also be considered, yet appears as 'beefy red' and macerated. The possibility of dependent edema was raised, given the patient's cardiovascular condition; however, this is rarely localized to such a small area and was summarily ruled out due to the clear demarcation of the discolouration.

Sepsis and septic shock are commonly seen in the paediatric population. One retrospective analysis (2) reported that 26.7% of total admissions to a paediatric intensive care unit were identified as septic shock. The attack rate of sepsis in full-term infants is 0.1% to 1%, and the associated mortality rate is roughly 20%.

Once the diagnosis of sepsis is established, aggressive treatment is necessary and life-saving. The causative organism should be sought, so that an appropriate treatment regimen can be started. Initial treatment should include boluses of isotonic crystalloid to regain hemodynamic stability. Laboratory studies should include base deficit, lactate concentration and venous oxygen saturation to identify the severity of shock and to monitor the response to fluid therapy. The patient should be monitored for metabolic derangements (eg, glucose and calcium), so that adjustments can be made. Finally, the need for vasopressors and stress-dose hydrocortisone should be anticipated to correct for adrenal insufficiency. In the event that severe shock persists, additional steps need to be taken (3).

CLINICAL PEARLS

- Assume a high degree of suspicion for sepsis in children.
- Not all suspected abuse is really abuse. Consider a broad differential when diagnosing perianal or vaginal discolourations. Most children referred for evaluation of sexual abuse have normal or nonspecific physical examinations.
- Aggressive treatment of septic shock can be life-saving.

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CASE 2 DIAGNOSIS: FREY (AURICULOTEMPORAL NERVE) SYNDROME

Localized unilateral or bilateral facial flushing in the auriculotemporal nerve (ATN) territory (lateral cheek, medial ear and frontotemporal scalp) is known as Frey syndrome. It is often triggered by masticulatory or tactile stimuli and may also be associated with hyperhidrosis in the area.

The syndrome was originally described by Duphenix in 1757, and later by Bailarger in 1853, who reported this in patients following parotid abscess surgery. The condition later gained its eponym in 1923 after Frey made the pathophysiological association with the ATN.

The ATN arises as two roots from the posterior division of the mandibular nerve. It emerges on the face behind the temporomandibular joint within the surface of the parotid gland and supplies somatosensory and parasympathetic innervation to the face. The somatosensory component of the auriculotemporal nerve innervates the skin overlying the preauricular areas, tragus, auricle and temporal region. The parasympathetic component carries postganglionic fibres to the parotid and sweat glands.

In the case of Frey syndrome, clinical findings are presumed to be due to injury or aberration associated with the autonomic component of the ATN. During healing, nerve fibres intended for the parotid may be misdirected and feed the sympathetic nerve fibres, which innervate the sweat glands and small blood vessels, explaining the erythema and hyperhidrosis seen in response to gustatory stimuli.

In adults, Frey syndrome is most commonly seen following obvious trauma such as surgery due to parotid or cerebellopontine angle tumour disease, cervical sympathectomies or radical neck dissections. In children, the incidence of Frey syndrome is low, but this is likely due to under-reporting and under-recognition of cases.

In some cases, there is history of local trauma in the parotid area, most commonly at birth, although reports of accidental trauma later in life causing Frey syndrome have been reported.

Frey syndrome usually manifests in early infancy following exposure to solid foods. Vigorous chewing elicits a stronger stimulation of the parotid glands. The erythema typically begins shortly after mastication of food and lasts for 15 min to 45 min. It is not accompanied by discomfort, itching or burning. In contrast to adults, hyperhidrosis does not usually occur in children with Frey syndrome, possibly due to the immaturity of their sweat glands.

Because of the development of symptoms after eating, many patients are initially suspected of having a food allergy. Frey syndrome may be mistaken for food-induced allergic cutaneous reactions such as oral allergy syndrome, acute urticaria and allergic contact dermatitis. However, there are distinguishing features.

In oral allergy syndrome, patients typically experience itching and swelling of the lips and face following ingestion of certain foods, including apples, peaches, nuts and certain raw vegetables. Acute urticaria and allergic contact dermatitis are also pruritic.

In Frey syndrome, the gustatory flushing is benign and nonprogressive. No specific treatments are necessary in affected children, and the flushing disappears spontaneously with time. However, recognition of the process is important to avoid mislabelling of children as having a food allergy or subjecting them to unnecessary laboratory testing.

CLINICAL PEARLS

- In cases of asymptomatic unilateral or bilateral facial flushing, consider the possibility of Frey syndrome.
- Frey syndrome may be mistaken for oral allergy syndrome, food-induced urticaria and allergic contact dermatitis.
- Prompt recognition of Frey syndrome will reduce unnecessary food elimination and diagnostic testing.

RECOMMENDED READING

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