

(Figure). She had slightly increased peripheral tone, fisting of both hands, and slightly increased reflexes bilaterally. A swallow study ruled out aspiration. Magnetic resonance imaging (MRI) of the brain was ordered due to increased tone and revealed no abnormal intracranial findings but did show asymmetric macroglossia. She was referred to otolaryngology, where flexible laryngoscopy ruled out intrinsic tongue pathology, and to genetics for evaluation for Beckwith–Wiedemann syndrome (BWS). Her alpha-fetoprotein (AFP) level was elevated at 184 ng/mL (normal, 0–77 ng/mL), and abdominal ultrasound at age 2 months demonstrated asymmetric kidneys (right kidney, 6.5 cm; left kidney, 5.3 cm). Methylation of IC1 and IC2 of the BWS locus on chromosome 11p15 was normal; however, genetic testing for BWS can be negative in up to 30% of affected cases. A diagnosis of BWS was made based on the major findings<sup>1</sup> of macroglossia and hemihyperplasia of a body part (her tongue).

BWS is a growth disorder with a prevalence of 1 in 10 000 and no clear sex or ethnic predilection. The most common mechanism of inheritance is due to epigenetic imprinting defects in the BWS locus on chromosome 11p15. Its presentation can be highly variable, with characteristic findings including hemihypertrophy, macrosomia (present in ~50%), macroglossia (present in ~90%), omphalocele, cardiomyopathy and long QT syndrome, neonatal hypoglycemia, renal abnormalities, organ enlargement, and various neoplasms, including rhabdomyosarcoma, neuroblastoma, hepatoblastoma, and Wilms tumor. Difficult deliveries are common owing to macrosomia. In some infants, such as this child, the clinical findings may be less obvious. For infants with cardinal features of BWS but negative genetic testing, the reported incidence of tumors in early childhood is as high as 6.2%.<sup>2</sup> Surveillance screening with an abdominal ultrasound every 3 months during the first 8 years of life and serum AFP every 3 months for the first 4 years is the standard of care.

Multiple serious conditions present with macroglossia, including glycogen storage disease, lysosomal storage disorders, and BWS, all of which have treatment and screening guidelines. The macroglossia for this case is asymmetric and non-progressive, which supports the diagnosis of BWS. Macroglossia should raise concern for BWS even in the absence of more obvious manifestations, such as large size for gestational age or organomegaly, to ensure appropriate clinical screening and follow-up. ■

## Data Statement

Data Sharing Statement available at [www.jpeds.com](http://www.jpeds.com).

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## Sequential Retinal Hemorrhages in an Asymptomatic Child



An asymptomatic 4-year-old girl with an unremarkable medical history was seen in the pediatric ophthalmology clinic after failing her school vision test. On examination, her visual acuity was 20/32 and 20/25 in right and left eyes, respectively. The left retina appeared healthy, and the right macula showed intraretinal hemorrhage. One week later, her right acuity decreased to 20/125. Widefield fundus photography showed multiple white-centered hemorrhages with tortuous retinal vessels in both eyes (Figure). A systemic cause was suspected because the hemorrhages occurred bilaterally. Examination revealed gross splenomegaly. Blood test showed a white cell count of

460 000 per cubic millimeter (reference range, 5000–17 000) with a neutrophil count of 357 000 per cubic millimeter (reference range, 1000–8500). Subsequent blood film and bone marrow aspiration confirmed the diagnosis of chronic myeloid leukemia. The patient was treated with imatinib and hydroxycarbamide to which she responded well, with white cell count returning to normal level and spleen no longer palpable within 1 month of treatment.

Differential diagnoses for white-centered retinal hemorrhages, or Roth spots, in a child include subacute bacterial endocarditis, leukemia, anemia, anoxia (such as from carbon monoxide poisoning), intracranial hemorrhage from arteriovenous malformation, HIV microangiopathy, and dysproteinemia.<sup>1,2</sup> Initial investigations should be guided by thorough history and examination. For suspected bacterial endocarditis,



**Figure.** Widefield fundus photography showed multiple white-centered hemorrhages bilaterally, with tortuous retinal vessels.

blood tests should include complete blood count, erythrocyte sedimentation rate, C-reactive protein, and blood culture. An echocardiogram should also be arranged to look for vegetation. Complete blood count would also be helpful in identifying anemia and leukemia, as in this case. Lastly, HIV antibody testing should be performed in suspected cases. ■

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## Peritonsillar Abscess in an Infant



**A**n 8-month-old girl presented to the emergency department (ED) with a 3-day history of fever, decreased oral intake, and limited neck and jaw movement. Her examination was notable for right-sided palatal edema, a left-shifted uvula, and an obstructed view of the oropharynx. A computed tomography (CT) scan of the neck demonstrated a 4-cm right peritonsillar abscess with significant mass effect on the airway (**Figure 1**). She was managed with noninvasive airway support and intravenous steroids plus intravenous ampicillin/sulbactam. She was taken to the operating room by the otolaryngology service, who drained 12 mL of purulent fluid from the abscess. She rapidly improved and was discharged from the hospital on day 4 with a course of oral amoxicillin/clavulanic acid.

Peritonsillar abscess is a suppurative tissue infection that occurs in the palatine tonsils. The presentation includes fever, dysphagia, drooling, and trismus. It is a rare condition

in children younger than 5 years of age, with a mean age of onset of 12 years and two-thirds of the cases occurring in children aged >10 years.<sup>1</sup> Sepsis, jugular vein thrombosis, and airway obstruction are potential serious complications.<sup>2</sup> Smaller infant airways may be more susceptible to compromise, as evidenced in this patient, and thus more difficult to support (noninvasively or invasively) in critical scenarios.

As in this case, fever and decreased oral intake may be the only initial complaints in an infant with a peritonsillar abscess. These common pediatric symptoms are nonspecific, making a peritonsillar abscess difficult to diagnose, especially if clear visualization of the oropharynx is not performed or if these symptoms are attributed to another more common infectious etiology in this age group, such as Coxsackievirus or primary herpetic gingivostomatitis. Careful physical examination, including good visualization of the oropharynx, is key to suspecting the diagnosis of a peritonsillar abscess in an infant. Recognizing the vulnerability of the infant airway in the setting of a deep space neck infection is paramount to supporting the airway and