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## Visual Diagnosis : A 5-year-old Child Who Has Facial Palsy and Rash

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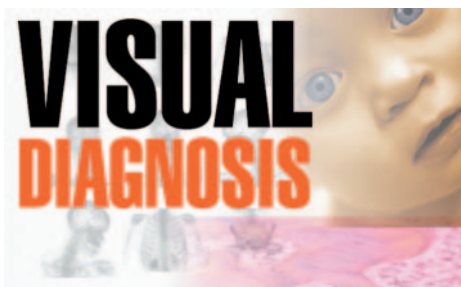
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## A 5-year-old Child Who Has Facial Palsy and Rash

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### Presentation

A 5-year-old girl who has a history of eczema and seasonal allergic rhinitis presents to the pediatric emergency department with the complaints of an enlarging rash and right-sided facial weakness. The patient was seen by her pediatrician 5 days ago and was diagnosed with a flulike illness. Her pediatrician recommended ibuprofen for relief of her symptoms. Two days ago, her mother noticed a rash on the girl's right leg that has expanded in size. On awakening this morning, she was unable to move the right side of her face. No other family members are ill.

Physical examination reveals a well-appearing child in no acute distress. Her weight is normal for age, and vital signs are within normal limits. Facial asymmetry is evident, with right-sided facial weakness and a droop in the right corner of her mouth (Figure). She cannot wrinkle her forehead or close her eye completely on the affected side. There is no nuchal rigidity, and Kernig and Brudzinkski signs are normal. A 5-cm round, symmetric, blanching erythematous rash with central clearing that is slightly warm is apparent on her right anterior thigh. There are no other rashes, and the remainder of the physical examination findings are normal. A brief video of her general examination is provided (**Click for Video**).

A complete blood count demonstrates a hematocrit of 38% (0.38) and a white blood cell count of  $5.6 \times 10^3/\text{mcL}$  ( $5.6 \times 10^9/\text{L}$ ) with 45% neutrophils, 27% lymphocytes, and 14% monocytes. The platelet count is  $274 \times 10^3/\text{mcL}$  ( $274 \times 10^9/\text{L}$ ). Serum electrolyte, alanine transaminase, and aspartate transaminase values are normal. The erythrocyte sedimentation rate is 24 mm/hr. Additional serum testing further supports the diagnosis after consideration of the child's environmental exposures and season of year.

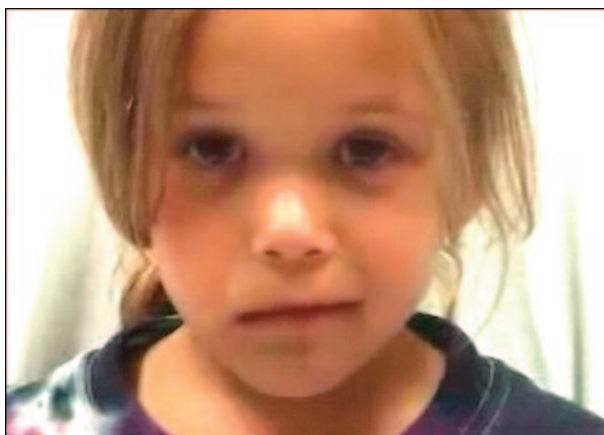


Figure. Right-sided facial weakness.

#### Author Disclosure

Drs Arnold and Spiro did not disclose any financial relationships relevant to this case.

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## Diagnosis: Lyme Disease With Erythema Migrans and Bell Palsy

An enzyme-linked immunosorbent assay (ELISA) for *Borrelia burgdorferi* is positive, and the result is confirmed by a Western blot test.

### Etiology, Epidemiology, and Definition

Erythema migrans (EM), the classic early manifestation of Lyme disease (LD), was described in the early 20th century by Afzelius. Epidemiologic investigation of an apparent case cluster of juvenile rheumatoid arthritis in Lyme, Connecticut, in 1975 led to the identification of LD as a distinct disease. The spirochete *B burgdorferi* was detected in 1981 as the cause of this tick-borne, multisystem inflammatory illness. Of the species in the genus *Borrelia* that cause LD, *B burgdorferi* is the sole cause in the United States (In Europe and Asia, most cases of LD are due to *B burgdorferi*, *B afzelii*, or *B garinii*). Manifestations of LD in the United States may differ from those in Europe; this discussion is confined to LD as it occurs in the United States.

*Borrelia* live in reservoir-competent hosts (white-footed mice in the northeast and north central United States), from which they are transmitted to *Ixodes* (black-legged) ticks. There are four stages in the 2-year life cycle of the tick (egg, larva, nymph, and adult), with larvae and nymphs feeding on the white-footed mouse or other rodents and adults feeding primarily on deer. Humans become infected as incidental hosts after contact with infected ticks.

The Centers for Disease Control and Prevention has established a definition of LD for epidemiologic surveillance as “physician-diagnosed erythema migrans  $\geq 5$  cm in diameter or at least one objective manifestation of late LD with laboratory confirmation of *B burgdorferi* infection using a two-tiered assay.” However, this definition is too strict for clinical use, and adherence to it will miss some cases of LD. Although most cases occur in the northeast, mid-Atlantic, and north central states, LD was reported in all states except Montana, Hawaii, and Oklahoma in 2002. As a result of the activity of the larval and nymphal ticks during late spring and early summer, 78% of cases are reported between May and August. LD is the most common vector-borne illness in North America and Europe. However, it is rare in the southeastern United States because *Ixodes scapularis* primarily feed on lizards that are resistant to *B burgdorferi*.

### Clinical Features

Similar to syphilis, the other spirochetal disease of humans, LD progresses through three somewhat distinct stages that overlap and reflect localized entry and dissemination of the organism (Table 1). The first, localized stage is heralded by the characteristic slowly expanding skin lesion, known as EM, which often is described as a solid red rash that forms a ring or multiple rings, giving a “bull’s-eye” appearance. EM is present in 70% to 80% of cases, and the presence of EM alone is sufficient for diagnosing LD during this stage. Localized disease also

Table 1. Key Features in Untreated Patients and Diagnostic Strategies for Lyme Disease

Stage	Timeline	Clinical Features	Diagnostic Criteria	Ancillary Diagnostic Tests
1. Localized	3 to 32 days	EM (70% to 80%), flulike illness	History of possible <i>B burgdorferi</i> exposure plus EM	
2. Early Disseminated	Days to months	<ul style="list-style-type: none"> <li>• Arthritis (60% to 70%)</li> <li>• Neurologic (10%)</li> <li>• Carditis (8%)</li> <li>• Conduction defects</li> <li>• Myopericarditis</li> </ul>	History of possible <i>B burgdorferi</i> exposure plus clinical features	ELISA and, if positive, Western blot
3. Late Disseminated	Months to years	<ul style="list-style-type: none"> <li>• Arthritis (50%)</li> <li>• Arthralgia (20%)</li> <li>• Tertiary neuroborreliosis</li> <li>• Encephalopathy</li> <li>• Peripheral neuropathy</li> </ul>	History of possible <i>B burgdorferi</i> exposure plus clinical features	ELISA and, if positive, Western blot

EM=erythema migrans, ELISA=enzyme-linked immunosorbent assay.

may be accompanied by fatigue, headache, arthralgias, myalgias, and fever, as noted in this case.

Disseminated infection occurs primarily in untreated patients. Early disseminated infection occurs days to months after initial manifestations of the illness and may be the initial manifestation of LD. Secondary skin lesions, similar to the primary EM, may occur during early disseminated infection. Indeed, these lesions may be the most apparent manifestation of dissemination. In patients not yet treated with antibiotics, other signs and symptoms may include mono- or oligoarticular arthritis (60% to 70%), predominantly of large joints. Second in frequency is neurologic involvement (10%), usually Bell (CN VII) palsy that may be bilateral. In endemic areas, LD is included in the differential diagnosis for CN VII palsy.

Manifestations of late disseminated disease may ensue months to years after spirochete entry. The bacteria survive in localized areas but escape detection by the immune system. Manifestations of late disseminated disease may include tertiary neuroborreliosis, but migratory polyarthritis and chronic arthritis predominate.

The Infectious Diseases Society of America has published evidence-based guidelines for diagnosis and treatment of LD. Most patients respond to these recommended regimens. However, a small proportion of patients may have persistent synovitis. A second group of patients experience more debilitating musculoskeletal pain, cognitive deficits, or fatigue. Whether these clinical manifestations represent fibromyalgia or a true “chronic LD” is questionable because affected patients are as likely to respond to placebo as they are to antibiotic treatment.

## Diagnosis

The overall approach to diagnosis is based both on the likelihood of exposure to *B burgdorferi* and clinical findings. An important caveat is that LD does not occur in the absence of signs and symptoms; clinical signs and symptoms are essential for disease recognition. Indeed, the diagnosis is made most often in patients who possibly have been exposed to ticks and present with EM.

For a patient who has a possible exposure to *B burgdorferi*, EM alone warrants diagnosis and treatment. On the other hand, patients presenting with signs and symptoms of disseminated disease (both early and late) are more likely to manifest positive serology. In these patients, positive serology should be used to support the clinical impression of early or late disseminated disease.

Results of serologic testing may be confusing when diagnosing early disseminated or late LD. Most infections have a detectable immunoglobulin M (IgM) re-

sponse within 2 weeks of onset. However, in LD, such a response may take up to 4 weeks to occur. Furthermore, there are similar delays in IgG response (6 to 8 weeks).

ELISA testing has high sensitivity but low specificity. In areas of low prevalence, or in patients who have more nonspecific symptoms, a positive ELISA result is more likely to be falsely positive due to cross-reactivity with spirochetes in normal oral flora. For this reason, a test with greater specificity, the Western blot, is used to confirm a positive ELISA finding. Therefore, making the serologic diagnosis of early disseminated or late LD involves a two-tiered approach, with a positive ELISA result warranting confirmation by a Western blot. A negative ELISA result indicates a negative serologic diagnosis for LD and does not need to be confirmed by Western blot.

## Differential Diagnosis

The differential diagnosis for LD is broad due to the potential multisystem manifestations of the disease. Moreover, many common illnesses have symptoms similar to those of LD. Fatigue, headache, arthralgias, myalgias, and fever warrant consideration of influenza and other viral illnesses. This constellation of signs and symptoms should prompt consideration of LD, especially during the warm weather months in an endemic area.

Another differential diagnosis for LD is the southern tick-associated rash illness (STARI), which has been identified in the southeastern United States. STARI has features similar to LD, including mild influenzalike symptoms and an EM-like skin lesion. The Lone Star tick has been identified as the vector, but the infectious agent has yet to be isolated.

The differential diagnosis for EM includes erythema annulare, erythema marginatum, pityriasis rosea, subacute cutaneous lupus, syphilis, juvenile idiopathic arthritis, erysipelas (if central clearing does not occur), and tinea corporis. In particular, EM often is diagnosed as tinea corporis and vice versa.

The differential diagnoses for Lyme arthritis include pauciarticular juvenile idiopathic arthritis and reactive arthritis.

The cause of CN VII palsy is unknown in approximately 70% of cases. Identified causes, in addition to LD, include herpes simplex virus (HSV)-1, human immunodeficiency virus, and varicella zoster virus (VZV) infections; reactivation of HSV-1 or VZV (Ramsey Hunt Syndrome); and recent administration of the intranasal influenza vaccine.

**Table 2. Recommended Antimicrobials for Children Who Have Suspected Lyme Disease**

Indications	Antimicrobial	Dosage	Duration
Tick bite	Doxycycline PO	4 mg/kg $\geq$ 8 y of age only	One dose
Erythema migrans, Lyme arthritis, CN VII palsy (Bell palsy)	Doxycycline PO	4 mg/kg per day $\div$ BID (maximum, 100 mg/ dose)	14 days
	Amoxicillin PO	50 mg/kg per day $\div$ TID (maximum, 500 mg/ dose)	14 days
	Cefuroxime PO	30 mg/kg per day $\div$ BID (maximum, 500 mg/ dose)	14 days
Lyme meningitis	Ceftriaxone IV	50 mg/kg per day $\div$ QD	14 to 28 days
	Cefotaxime IV	150 mg/kg per day $\div$ TID	14 to 28 days

See: <http://www.journals.uchicago.edu/CID/journal/issues/v43n9/40897/40897.html> for comprehensive treatment guidelines.

### Treatment

The best method to prevent LD is to avoid exposure to the tick vector. Measures to reduce exposure and risk of infection include: the use of tick repellents, use of protective clothing, examination of the child's entire body for ticks, and immediate removal of an attached tick. For children 8 years of age and older, doxycycline (4 mg/kg up to 200 mg maximum daily dose) may be prescribed after a tick bite in an endemic area if all of the following conditions are present: 1) the tick is found to be engorged, 2) the tick has been attached for a minimum of 36 hours, 3) prophylaxis can be started within 3 days. Doxycycline is not preferred in children younger than 8 years of age because it can discolor teeth permanently. The data supporting the efficacy of antibiotic prophylaxis for children younger than 8 years of age are scarce.

Doxycycline (for children  $\geq$ 8 years of age), amoxicillin, or cefuroxime axetil for 14 days is recommended for children presenting with EM or Lyme arthritis in the absence of neurologic symptoms (Table 2). Macrolides such as erythromycin are less effective but may be used for children who have drug sensitivities to the previously noted agents. If bacterial cellulitis cannot be distinguished from EM, amoxicillin-clavulanate should be considered. For children who have Lyme meningitis, intravenous ceftriaxone in a single daily dose for 14 days is recommended. For patients who have an isolated CN VII palsy without evidence of Lyme meningitis, as in this case, a 14-day oral regimen may be used. Although cerebrospinal fluid pleocytosis is associated with CN VII palsy due to LD, routine lumbar puncture may not be necessary unless the clinician suspects meningitis on the basis of the symptoms and physical examination findings.

### Patient Course

The child was diagnosed clinically with early disseminated LD and treated with amoxicillin, 50 mg/kg per day in three divided doses for 14 days. Supportive measures to treat the CN VII palsy included lubricant eye drops and education and reassurance to both the child and the family. A lumbar puncture was not performed. The rash faded and resolved within 5 days of treatment with antibiotics, and the facial nerve paralysis resolved 2 weeks after the initial presentation to the ED.

### Conclusion

LD may have various clinical presentations, including EM and CN VII palsy, as in this patient. The diagnosis may be made clinically without laboratory testing in endemic areas. If the clinician suspects LD, the child should be treated presumptively. For children presenting with EM and isolated CN VII palsy without signs or symptoms of meningitis, a lumbar puncture may not be necessary.

### Suggested Reading

- Alberton DL ZP. Bell's palsy: a review of treatment using antiviral agents. *Ann Pharmacother*. 2006;40:1838–1842
- Lyme disease—United States, 2001–2002. *MMWR Morbid Mortal Wkly Rep*. 2004;53:365–369
- Stafford KC III. *Tick Management Handbook: An Integrated Guide for Homeowners, Pest Control Operators, and Public Health Officials for Prevention of Tick-Associated Disease*. New Haven, Conn: The Connecticut Agricultural Experiment Station; 2004



Stanek G, Strle F. Lyme borreliosis. *Lancet*. 2003;362:1639–1647

Steere AC. Lyme borreliosis in 2005, 30 years after initial observations in Lyme Connecticut. *Wien Klin Wochenschr*. 2006;118:625–633

Steere AC, Coburn J, Glickstein L. The emergence of Lyme disease. *J Clin Invest*. 2004;113:1093–1101

Weston WL LA, Morelli JG. *Color Textbook of Pediatric Dermatology*. St. Louis, Mo: Mosby; 2002

Wormser GP, Dattwyler RJ, Shapiro ED, et al. The clinical assessment, treatment, and prevention of Lyme disease, human granulocytic anaplasmosis, and babesiosis: clinical practice guidelines by the Infectious Diseases Society of America. *Clin Infect Dis*. 2006;43:1089–1134

## Clarifications

### Heat Illness and Heat Stroke (July 2007, pp 249–258)

Two readers have pointed out a discrepancy in this article. In the last paragraph on page 253, readers are advised to treat heat stroke by cooling the patient to less than 104°F (40°C), but Table 3 on page 254 recommends cooling the patient until the core temperature is less than 102.2°F (39°C). Neither the editors nor the author (Dr David Jardine) noted the inconsistency. According to Dr Jardine, “It is important that the patient be cooled to less than 40°C as rapidly as possible because cellular damage stops occurring once the temperature is below 41°C. It is not critical to lower the temperature rapidly to 39°C except in the case of severe heat stroke. In this situation, the patient may be intolerant of the additional physiologic stress caused by elevated body temperature.”

### Hypertension in Childhood (August 2007, pp 283–298)

Table 2 on page 286 lists figures that differ slightly from those found in “The Fourth Report on the Diagnosis, Evaluation, and Treatment of High Blood Pressure in Children and Adolescents” (*Pediatrics*. 2004;114:555–576). The author of the PIR article (Dr Leonard Feld) advises that the definition of Stage 1 hypertension in Table 2 should read: “Average SBP or DBP that is 95th to 99th percentile plus 5 mm Hg.” The definition of hypertensive urgency and emergency should read: “Average SBP or DBP that is >99th percentile plus 5 mm Hg, along with clinical signs or symptoms.” Dr Feld adds, “From my clinical practice point of view, I would be concerned with any child whose blood pressure exceeded the 95th percentile for age and height that was sustained.”

### Chronic Abdominal Pain (September 2007, pp 323–331)

On page 327, in the second sentence of the third paragraph of the section on IBS, the parenthetical phrase describing abnormal stool patterns should read: “(four or more stools per day [rather than per week] or <2 stools per week).”

### Index of Suspicion (October 2007, pp 389–394)

In Case 2 Presentation on page 389, the international units for the electrolyte values are incorrect. The correct units are: calcium concentration 4.475 mmol/L (normal, 2.05 to 2.675 mmol/L), ionized calcium concentration 2.05 mmol/L (normal, 1.125 to 1.325 mmol/L), and phosphorus concentration 0.71 mmol/L (normal, 0.84 to 1.62 mmol/L).

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