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# Congenital heart block in neonatal lupus erythematosus

NLE occurs in 1 out of every 15,000 live births annually and accounts for 85% of all cases of congenital heart block in the absence of cardiac structural abnormalities. Here's what you need to know.

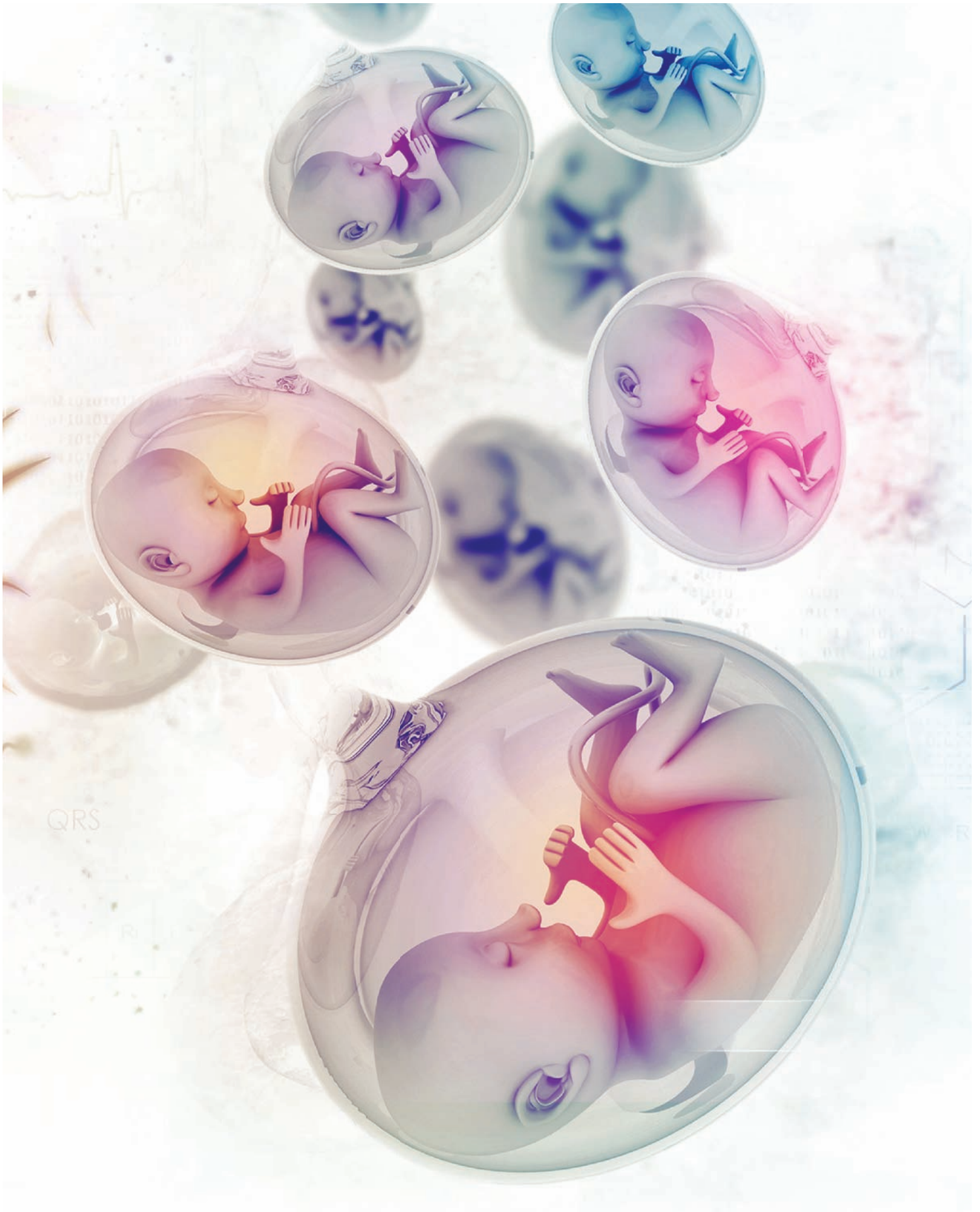
By Richard L. Pullen, Jr., EdD, MSN, RN, CMSRN, and Marietta Branson, DHSc, MSN, RNC

Neonatal lupus erythematosus (NLE) is a passively acquired autoimmune disease in which maternal autoantibodies against the antigens Sjögren syndrome A (SS-A/Ro), Sjögren syndrome B (SS-B/La), and ribonucleoprotein (RNP) lead to tissue damage in the fetus. NLE occurs in 3% of pregnant women with these autoantibodies, 1 out of every 15,000 live births, and 0.6 of every 100,000 children.

The most serious sequelae of NLE occur when autoantibodies attack the cardiocytes in the fetus during the second or

third trimesters, causing an immune-mediated congenital heart block (CHB), cardiomyopathy, valvular insufficiency, and heart failure. NLE accounts for 85% of all cases of CHB in the absence of cardiac structural abnormalities; 60% of infants and children require pacemakers and 20% die from heart failure. Large prospective studies suggest that most deaths from CHB caused by NLE occur in utero. Approximately 20% of infants with NLE at birth develop cardiomyopathy during the childhood years.

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In this article, we discuss the pathogenesis of CHB in NLE and management approaches for the mother, fetus, and infant.

### **Pathogenic mechanisms of autoantibodies**

The SS-A/Ro, SS-B/La, and RNP complex is a subgroup of antinuclear antibodies located within the nucleus of the cytoplasm and commonly found in patients with autoimmune disease. High titers of these autoantibodies in the mother place the fetus at risk for CHB; fetal surveillance by echocardiography is warranted. These autoantibodies may affect the sinoatrial and atrioventricular nodes, causing varying degrees of atrioventricular (AV) block. CHB is most often diagnosed in utero through routine ultrasound and confirmed by fetal echocardiography.

In addition to affecting the cardiac conduction system, maternal autoantibodies cause ventricular dilatation and myocardial hypertrophy and induce endocardial fibroelastosis (thickening of the ventricular myocardium), frequently leading to end-stage heart failure and death. Fetal hydrops, in which there's an excessive accumulation of fluid in two or more systems or compartments, such as pericardial and pleural effusions and ascites, is also a complication with high mortality.

SS-A/Ro is present in 40% of patients with systemic lupus erythematosus (SLE) and 70% of patients with Sjögren syndrome, triggering a plethora of clinical syndromes, including complete AV block, a prolonged QT interval, life-threatening dysrhythmias, photosensitive skin rashes, nephritis, and glandular and extraglandular symptoms. SS-A/Ro is also implicated in at least 95% of cases of CHB caused by NLE. There are several theories why this antibody is pathogenic in CHB.

First, SS-A/Ro includes two specific proteins that are highly pathogenic: 52 kD Ro and 60 kD Ro. These maternal

autoantibodies enter the fetal circulation through the placenta and bind to cardiocytes. Reactivity to the p200 antigenic component of 52 kD Ro increases the risk of third-degree AV block. The deposition of SS-A/Ro in cardiocytes stimulates the production of macrophages and proinflammatory cytokines, such as tumor necrosis factor alpha and transforming growth factor beta, which leads to inflammation, tissue damage, apoptosis, and fibrosis.

Second, the L-type calcium channel is a conduction pathway between the atria and ventricles. SS-A/Ro inhibits conduction through this pathway, causing calcium dysregulation in cardiocytes. Dysregulation prolongs the exposure of the L-type calcium channel to SS-A/Ro and leads to apoptosis and fibrosis.

Third, SS-A/Ro is strongly associated with conduction disturbances in the fetus because it blocks the 5-hydroxytryptamine (serotonin) receptor 4, which normally causes cardiac stimulation through the release of norepinephrine from sympathetic nerve channels.

### **Maternal risk factors**

Approximately 60% of women have an autoimmune disease when NLE is diagnosed, whereas 40% of women don't appear to have an underlying disease. Women with known autoimmune disease should already be under the care of a rheumatologist and obstetrician with high-risk pregnancy experience who are familiar with her clinical status before pregnancy and in the perinatal period. The patient's SS-A/Ro titers should be closely evaluated, as well as the presence of other autoantibodies suggestive of autoimmune disease, including antiphospholipid antibodies (thrombosis), antidouble stranded DNA (renal disease in SLE), rheumatoid factor (rheumatoid arthritis), and thyroid peroxidase (Hashimoto thyroiditis).

In women who have no known disease, the presence of maternal autoantibodies may be noted when the fetal heart rate begins to decrease and fetal conduction deficits emerge. This may be the first indication that the woman has an autoimmune disease. A serum blood test may confirm the presence of autoantibodies. Diagnosing an infant with NLE doesn't always predict that the mother will have an autoimmune disease. However, women who are asymptomatic may develop clinically significant disease within 10 years of delivering an infant with NLE. The mother may genetically predispose the fetus to CHB if she also possesses the human leukocyte antigen-B8 (HLA-B8) and human leukocyte antigen-DR3 (HLA-DR3). Once a woman with SS-A/Ro gives birth to an infant with CHB, she has a 15% risk of having another infant with CHB. The risk of CHB increases in gestational weeks 18 to 24. Fetal sex doesn't appear to be a significant risk factor for CHB.

### Fetal surveillance

Fetal monitoring through M-mode pulse Doppler echocardiography can identify early CHB and evaluate cardiac rate, rhythm, and function. This form of echocardiography measures synchrony between the mitral valve inflow and aortic valve outflow from the ventricular outflow tract. The difference in time delay between mitral valve inflow and aortic valve outflow relates to atrial and ventricular systole to establish a mechanical PR interval.

Fetal magnetocardiography is a non-invasive procedure used to evaluate the progression of CHB that uses magnetic waves initiated by the fetal heart. This is a safe procedure similar to an ECG.

There's often an organized progression in the fetus from first-degree AV block, which is early onset and reversible, to second-degree AV block, and then third-degree AV block, which is irreversible. Approximately 30% of fetuses exposed

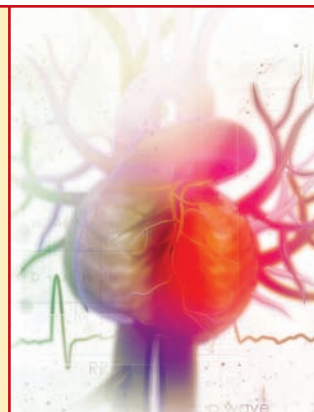
to SS-A/Ro have abnormal AV conduction, most becoming normal by delivery, whereas at least 5% progress to advanced AV block. The evidence suggests that most fetuses exposed to SS-A/Ro not only have conduction deficits, but also problems with systolic cardiac performance.

### Treating the mother and fetus

The mother should be closely monitored for disease exacerbations during pregnancy. For example, hypertension in pregnancy is common with SLE, which can lead to severe preeclampsia and renal failure. High titers of SS-A/Ro in a symptomatic or asymptomatic mother may correlate with the presence of additional autoantibodies and other lab markers of systemic inflammation.

Abnormally low levels of complement components 3 and 4 are reliable indicators of maternal disease activity because

The risk of congenital heart block increases in gestational weeks 18 to 24.

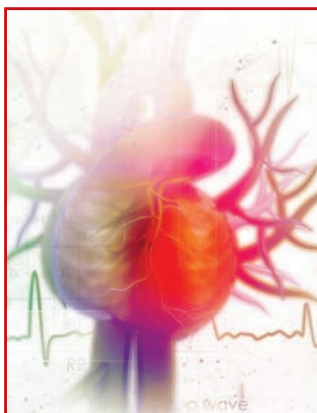


they're consumed in chronic inflammation. Complement are proteins produced by the liver in response to inflammation to clear the body of autoantibodies. High levels of C-reactive protein and erythrocyte sedimentation rate are also indications of inflammation.

Maternal inflammation is usually controlled by anti-inflammatory medications, including those that suppress the immune system. A therapeutic outcome

of immunosuppression is eliminating autoantibodies and improving symptoms in the mother and cardiac function in the fetus.

A fetus with CHB may have a heart rate of less than 55 beats/minute (normal: 110 to 160 beats/minute). Maternal immunosuppression with fluorinated steroids, such as dexamethasone and betamethasone, often reverses first- and second-degree AV block because they're only partially metabolized by the placenta and the active form



Pacemaker insertion in the neonate after delivery may be necessary if treatment with fluorinated steroids is ineffective.

of the medication reaches the fetus. Steroids, such as prednisone, prednisolone, and methylprednisolone, aren't effective because they're metabolized by the placenta.

A third-degree AV block isn't considered reversible. Although fluorinated steroids don't reverse third-degree AV block, they're sometimes effective in reducing the risks of developing cardiomyopathy and fetal hydrops. If the fetal heart alternates between second- and third-degree AV block, then fluorinated steroids are continued through delivery.

After delivery, the mother is slowly tapered from the steroid. Standardized treatment for a fetus sustaining a heart rate of 55 beats/minute or less is to combine fluorinated steroids with the beta-adrenergic receptor agonist terbutaline to improve cardiac conduction and increase the heart rate. Pacemaker

insertion in the neonate after delivery may be necessary if treatment with fluorinated steroids is ineffective.

The use of steroids creates risks for the mother, including infection, hypertension, diabetes, premature rupture of membranes, preterm labor, and preeclampsia. Maternal use of steroids may also affect the fetus, causing adrenal suppression and oligohydramnios (deficient amniotic fluid), which can lead to umbilical cord compression, fetal growth restriction, musculoskeletal deformities, and pulmonary complications. One recommendation is to limit the use of fluorinated steroids to 10 weeks to avoid these complications. The prophylactic use of fluorinated steroids in women who have a titer for SS-A/Ro and are pregnant or plan to become pregnant isn't recommended because of potential adverse reactions.

I.V. immunoglobulins (IVIG) in the mother may reduce inflammation and subsequent fibrosis in the fetal heart by neutralizing or eliminating pathogenic autoantibodies. Combining IVIG and fluorinated steroids with weekly plasmapheresis to reduce high titers of SS-A/Ro may stabilize a second-degree AV block and prevent the development of a third-degree AV block. Postdelivery treatment with fluorinated steroids alone or with IVIG isn't effective in managing CHB. However, infants with rare severe hepatic and hematologic manifestations may be treated with corticosteroids, such as methylprednisolone, other immunosuppressive medications, and IVIG.

Hydroxychloroquine is an antimalarial agent and disease-modifying antirheumatic drug that's commonly prescribed to reduce inflammation in SLE and other autoimmune diseases. It inhibits the production of proinflammatory cytokines to reduce inflammation. Longitudinal studies of infants whose mothers received hydroxychloroquine during pregnancy and breastfeeding suggest that it's safe for

the fetus and infant. These infants had normal growth and development. Standard treatment is hydroxychloroquine prophylaxis in women with SS-A/Ro before the development of signs and symptoms of CHB in the fetus. Visual acuity should be assessed in the mother and infant because hydroxychloroquine can rarely cause retinal damage. However, any damage from hydroxychloroquine is reversible once it's discontinued.

Early recognition and treatment of CHB, especially with fluorinated steroids, are crucial. In a multicenter study of in utero fluorinated steroid administration, 100% of fetuses with first-degree AV block converted to a normal sinus rhythm; 30% of infants with second-degree AV block converted to a normal sinus rhythm, 50% remained in second-degree block, and 20% progressed to third-degree AV block. One hundred percent of fetuses with third-degree AV block remained in third-degree block. In another study, 100% of fetuses with second-degree AV block treated with fluorinated steroids converted to first-degree AV block or better by delivery, whereas fetuses not treated progressed to third-degree AV block.

### Criteria for permanent pacemakers in infants

Assessment of cardiac function is essential once the neonate's respiratory rate, depth, and effort are stabilized upon delivery. An ECG should be performed in a timely manner to determine the extent of CHB. Cardiac involvement can be from mild to life-threatening upon delivery, and a pacemaker may be necessary. The neonate may initially present with symptomatic bradycardia and then progress to advanced forms of AV block (see *Types of heart blocks in NLE*). The child will need continued surveillance throughout his or her life.

A permanent pacemaker becomes necessary when there are symptoms related to bradycardia or second- or third-degree

## Types of heart blocks in NLE

### Sinus bradycardia with first-degree AV block

PR interval >0.20 s (Normal: 0.12 to 0.20 s)



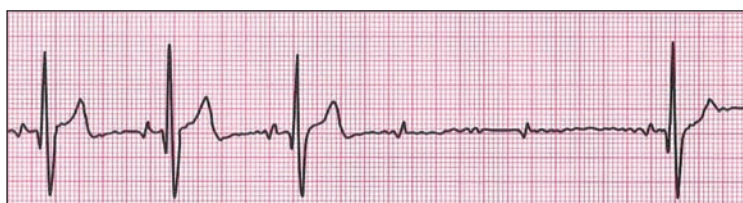
### Second-degree AV block Mobitz I (Wenckebach)

P-P interval consistent. Gradual prolongation of the PR interval until a P wave isn't followed by a QRS complex, making the rhythm irregular.



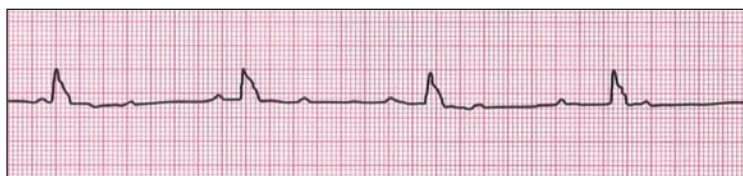
### Second-degree AV block Mobitz II

The PR interval remains consistent. P waves "march out" in a consistent manner. Some P waves are blocked before they reach the ventricle. A 2:1 block is noted when every other P wave is conducted.



### Third-degree AV block

No relationship between P waves and QRS complexes. Atrial impulses don't reach the ventricles. Ventricular rhythm is maintained by a slower pacemaker at the AV node.



Source: Adapted from American Heart Association, American Academy of Pediatrics. *Pediatric Advanced Life Support Provider Manual*. Dallas, TX: American Heart Association; 2017.

AV block, especially when an infant has diminished cardiac output. For the American College of Cardiology, American Heart Association, and European Society of Cardiology guidelines for pacemaker insertion in infants, see *Criteria for permanent pacemaker placement in infants*. A permanent pacemaker isn't indicated when an infant has third-degree AV block with an acceptable heart rate, ventricular function, narrow QRS complex, and sufficient cardiac output. Infants with asymptomatic type I second-degree AV block may not require a permanent pacemaker when there's an acceptable heart rate and sufficient cardiac output. Delayed CHB and cardiomyopathy may occur in infants, children, and adults, and should be closely monitored by a cardiologist throughout their lifetime.

### Clinical correlation of cardiac and noncardiac manifestations

Cardiac rate and rhythm in the fetus and neonate must always be correlated with hemodynamic status and responses to intrauterine and postdelivery treatment. Maternal, fetal, and neonatal surveillance

must also be initiated to evaluate adverse reactions of steroid therapy. The stress on the mother from a complicated pregnancy needs to be evaluated, as does its impact on comorbidities, such as SLE or Sjögren syndrome.

Cutaneous, hepatic, neurologic, and hematologic symptoms may coexist with or precede CHB and usually resolve when maternal autoantibodies have cleared the fetal circulation. The noncardiac manifestations of NLE are directly related to the inflammatory effects of SS-A/Ro on the skin, blood, liver, and central nervous system. Clinical correlation of noncardiac manifestations and the ECG must always be a priority.

Photosensitive, erythematous annular lesions with a hypopigmented center may appear on the head, periorbital area, face, and neck of the neonate. Lesions may have the histologic features of subacute cutaneous lupus erythematosus that resemble discoid lupus erythematosus. The lights in the delivery room, newborn nursery, and neonatal ICU should be dimmed to prevent the onset or exacerbation of lesions. The mother needs instruction about keeping the infant's room dimly lit, avoiding exposure to the sun, wearing protective clothing, using sunscreen with an SPF of at least 30, and applying topical corticosteroids as prescribed. The mother must also protect her skin in the same manner because she carries SS-A/Ro.

Hepatocytes may react to SS-A/Ro with elevated alanine aminotransferase and aspartate aminotransferase levels. The neonate may present with hepatitis and need to be treated with corticosteroids. Gamma-glutamyl transferase elevation may also indicate cholestasis, leading to jaundice in the infant. Serum albumin should be closely monitored because it's a reliable indicator of hepatic function.

Neutropenia, anemia, and thrombocytopenia may occur, and are usually mild and

#### Criteria for permanent pacemaker placement in infants

- Bradycardia reducing cardiac output due to sinus node dysfunction and a ventricular rate of 55 beats/minute or less
- Congenital advanced second- or third-degree AV block with a ventricular rate of 55 beats/minute or less
- Congenital third-degree AV block with ventricular ectopy
- Congenital third-degree AV block with ventricular dysfunction
- Congenital advanced second- or third-degree AV block with a ventricular rate of 70 beats/minute or less in the presence of congenital heart disease
- Prolonged QT-interval or wide QRS escape rhythm
- Post-op advanced second- or third-degree AV block that persists longer than 7 days

Sources: Adapted from Epstein AE, DiMarco JP, Ellenbogen KA, et al. ACC/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (writing committee to revise the ACC/AHA/NASPE 2002 guideline update for implantation of cardiac pacemakers and antiarrhythmia devices) developed in collaboration with the American Association for Thoracic Surgery and Society of Thoracic Surgeons. *J Am Coll Cardiol*. 2008; 51(21):e1-e62.

Nasef N, Hafez M, Bakr A. Neonatal lupus erythematosus. *J Neonatol Clin Pediatr*. 2014;1(002):1-10.

not treated. A complete blood cell (CBC) count and conjugated bilirubin levels must be closely evaluated for hemolytic anemia. Despite neutropenia, the evidence suggests that infection and sepsis don't usually occur. Persistent anemia and thrombocytopenia may be treated with IVIG and corticosteroids.

Most infants with NLE are born neurologically intact. One neuroimaging finding in NLE is echogenic lenticulostriate vasculopathy—streaks or spots in the thalamus and basal ganglia that may injure the fetal brain. This may predispose the infant to transient hydrocephalus, seizures, or vascular myelopathy. The infant's posture and muscle tone, primitive reflexes, and Babinski reflex should be assessed. Infants should be monitored periodically by cerebral sonography. Children should also be monitored for neuropsychiatric and behavioral problems and attention deficit disorder.

### Caring for the mother, fetus, and infant

Careful assessment of the mother, fetus, and infant during the perinatal period is critical when there's a diagnosis of CHB in NLE. You need to know what signs and symptoms are important indicators of potential cardiac problems in the fetus and infant. Implement priority nursing interventions in collaboration with the interprofessional team to decrease morbidity and mortality (see *Priority assessments for mother, fetus, and neonate*). The interprofessional team will need to follow the infant through his or her childhood and into adulthood to assess general health in relation to an implanted permanent pacemaker and cardiac status for the onset of delayed CHB and/or cardiomyopathy.

Most infants with or without a permanent pacemaker have a healthy and productive life. Mortality is 20% for infants with cardiomyopathy or a history of fetal hydrops. Some studies suggest that mortality is higher in non-White mothers of infants with NLE. There have been no

## Priority assessments for mother, fetus, and neonate

### Maternal assessments

- Assess:
  - history of lupus or other related autoimmune disease
  - SS-A/Ro, SS-B/La, and RNP titers
  - knowledge of genetic testing for HLA-B8 and HLA-DR3
  - decrease in complement levels, indicating autoimmune disease exacerbation
  - elevated sedimentation rate and C-reactive protein levels, indicating autoimmune disease exacerbation
  - hypertension, severe preeclampsia, diabetes, preterm labor, and premature rupture of membranes
  - infection during labor.
- Protect the mother from light sources because she's photosensitive due to the presence of SS-A/Ro.

### Fetal assessments

- Assess:
  - cardiac rate and rhythm at 18 to 24 weeks' gestation using ultrasound and Doppler echocardiography, and more often if CHB is noted
  - heart rate with continuous fetal monitoring during labor
  - bradycardia without cause
  - type of fetal heart block (first-, second-, or third-degree)
  - response to steroid administration.

### Neonatal assessments

- Assess:
  - respiratory status at delivery (rate, depth, and effort)
  - cardiac pattern (rate, rhythm, and pattern with auscultation and ECG)
  - heart block or symptomatic or asymptomatic bradycardia
  - the need for a pacemaker as determined by the degree of heart block
  - the skin for lesions
  - neurologic status
  - CBC count, bilirubin, and liver enzymes.
- Protect the neonate from light sources.
- Be on the alert for complications related to intrauterine steroids.

### Additional points

- Assess:
  - the parents for education and resource needs pertaining to cardiac NLE while in the neonatal ICU
  - support from other interprofessional team members such as counseling.
- Provide emotional support to the parents and assess for anxiety and fear of the unknown.
- Infants with permanent pacemakers need ongoing follow-up with a cardiologist through childhood and adulthood even when CHB was corrected in utero.
- The infant will be exposed to SS-A/Ro the longer the mother breast-feeds. Autoantibodies are passed from mother to infant in breast milk.





## on the web

### National Institute of Arthritis and Musculoskeletal and Skin Diseases:

[www.neonatalupus.com/neonatal-lupus.html](http://www.neonatalupus.com/neonatal-lupus.html)

### National Organization for Rare Disorders:

<https://rarediseases.org/rare-diseases/neonatal-lupus>

### The Johns Hopkins Lupus Center:

[www.hopkinslupus.org/lupus-info/lifestyle-additional-information/lupus-pregnancy](http://www.hopkinslupus.org/lupus-info/lifestyle-additional-information/lupus-pregnancy)

conclusive studies to suggest that infants with NLE will develop another autoimmune disease in their lifetime.

## Future considerations

Further longitudinal research is needed to follow children and adults of mothers with SS-A/Ro for the development of late-onset CHB and cardiomyopathy. There's also a need for nurses who specialize in the management of maternal, infant, and child care to enhance their knowledge and develop practice guidelines for patients and family members coping with NLE and other autoimmune diseases and syndromes that may impact quality of life. ■

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