

Pediatric Sjögren's

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Sjögren's is a systemic autoimmune disease that affects the entire body. Along with symptoms of extensive dryness, other serious complications include profound fatigue, chronic pain, major organ involvement, neuropathies, and lymphomas. This disease in children often presents differently than it does in adults; **the single most common presenting feature of childhood Sjögren's is swelling of the parotid gland(s) (with or without tenderness)**. The parotid gland is the major salivary gland(s) located over the angle of the jaw on both sides.

Sjögren's may cause:

- Dryness of eyes and mouth
- Joint pain (especially in the morning) or joint inflammation may occur and may be the initial manifestation
- Autoimmune attack on any type of blood cells may occur resulting in decreases in platelets, red or white blood cells
- Effects on kidneys may result in low potassium that may progress to paralysis. This is a medical emergency
- Inflammation of the optic nerve (the nerve that connects the eyes to the brain) and the spinal cord may manifest as blindness and paralysis. This condition, called *neuromyelitis optica*, is a medical emergency
- Effects on the lungs may include inflammation and pulmonary hypertension (increased blood pressure in the arteries within the lungs)

Sjögren's should be considered in any child with:

- Oral or ocular manifestations suggesting decreased saliva and/or tear production
- Swelling of parotid glands (persistent or recurrent)
- Inflammation of kidneys, lungs, brain, or spinal cord
- Autoimmune manifestations affecting the blood
- Symptoms including prolonged or recurrent fevers, enlarged lymph nodes, or a purple or reddish rash due to inflamed blood vessels or cold-induced color changes (white, blue, and/or red) of fingers or toes

Tests for Sjögren's may include blood tests, imaging studies, ocular and salivary gland tests, and a biopsy of the minor salivary glands located in the inner side of the lower lip.

Regular oral and eye health care is absolutely necessary for maximizing oral and ocular health. Additional management of pediatric Sjögren's is dictated by the specific symptoms of their disease. Potential strategies include:

- Short course of oral corticosteroids (e.g. prednisone) may help to improve symptoms short-term but additional therapy should be used to prevent long-term exposure to high doses of these medications.
- Typically, recurrent parotid gland swelling in children is treated with hydroxychloroquine (a daily oral medication) which often helps minimize episodes of recurrent parotid swelling and may be helpful with improving fatigue and joint pain often seen in pediatric Sjögren's.
- More potent immunosuppressive medications commonly used for other rheumatic diseases (including methotrexate, mycophenolate mofetil, azathioprine, rituximab, belimumab, abatacept) may be used especially if children have other organ involvement – including brain and spinal cord, lungs, joints, and blood.
- Children with Sjögren's should have regular visits with their Pediatric Rheumatologist, Ophthalmologist (eye doctor), and Dentist! These visits can help ensure good quality of life by preventing damage and monitoring for potentially serious complications.

For more information on Sjögren's, visit the Foundation website at www.sjogrens.org, call 301-530-4420, email info@sjogrens.org, or write to the Sjögren's Foundation, 10701 Parkridge Blvd, Ste 170, Reston, VA 20191