Medical Policy

Blue Cross Blue Shield Blue Care Network of Michigan

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Category: Laboratory/Pathology
***Current Policy Effective Date: 11/1/09**

Title: Urine Sulfate

**Procedure Code(s): 84392

Description / Background

This laboratory test may be ordered for the purpose of diagnosing certain abnormalities of sulfur metabolism. Sulfates may be measured for the diagnosis of metachromatic leukodystrophy (sulfatide lipidosis), an inherited lipid metabolism disorder that results in the accumulation of metachromatic lipids in the tissues of the central nervous system, leading to paralysis and often death in early adolescence. The specimen is a random or timed urine collection. The testing method used is spectrophotometry.

This test is performed to detect a rare disorder of excess acid mucopolysaccharides accumulation (especially dermatan sulfate and heparan sulfate) in the tissues. The severe form of this disorder can lead to Hurler syndrome. Typically this test is only ordered for infants when there is a family history of mucopolysaccharidosis (MPS).

Persons with Hurler syndrome do not make a substance called lysosomal alpha-L-iduronidase. This substance, called an enzyme, helps speed the break down of long chains of sugar molecules called glycosaminoglycans (formerly called mucopolysaccharides). These molecules are found throughout the body, often in mucus and in fluid around the joints. Without the enzyme, glycosaminoglycans build up and damage organs, including the heart. Symptoms can range from mild to severe.

Hurler syndrome is inherited autosomal recessive condition, which means that both parents need to pass down the faulty gene in order for the child to develop Hurler syndrome. Hurler syndrome

*See policy history boxes for any previous effective dates if applicable
**See section "CPT/HCPCS Level II Codes and Description" for code nomenclature and for additional code(s) if applicable.

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is a type of mucopolysaccharidosis called MPS I. Hurler syndrome is the most severe type. It is categorized as MPS I H. The other subtypes of MPS I are:

- MPS I H-S (Hurler-Scheie syndrome)
- MPS I S (Scheie syndrome)

Symptoms of Hurler syndrome most often appear between ages three and eight. Infants with severe Hurler syndrome appear normal at birth. Facial symptoms may become more noticeable during the first two years of life. Symptoms include:

- Thick, coarse facial features with a low nasal bridge
- Progressive mental retardation
- Deafness
- Joint disease, including stiffness
- Heart value problems
- Abnormal bones in the spine
- Claw hand

A bone marrow transplant may improve some of the symptoms of the disease. To prevent mental retardation, a bone marrow transplant probably should be done at a very young age. Other treatments depend on the organs that are affected.

CPT / HCPCS Level II Codes and Description *(Note: The inclusion of a code in this list is not a guarantee of coverage. Please refer to the medical policy statement to determine the status of a given procedure)*

Established codes:

84392 Sulfate, urine

Other codes (investigational, not medically necessary, etc.):

N/A

Diagnoses / Medical Conditions

- Leukodystrophy

Medical Policy Statement

The safety and effectiveness of urine sulfate testing have been established. It may be considered a useful diagnostic tool for diagnosing certain disorders of sulfur metabolism that cause excess acid mucopolysaccharide accumulation in the body.

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Rationale

Urine sulfate testing has been shown to be effective in diagnosing various medical conditions, including mucopolysaccharidosis. This code has been in effect and payable by Medicare and Medicaid since at least 2001.

Medical Policy Position Summary (Non-clinical summary statement for customer use)

Urine sulfate testing is performed to detect a rare disorder of excess acid mucopolysaccharides accumulation (especially dermatan sulfate and heparan sulfate) in the tissues. The severe form of this disorder can lead to Hurler syndrome. Typically this test is only ordered for infants when there is a family history of mucopolysaccharidosis (MPS).

Persons with Hurler syndrome do not make a substance called lysosomal alpha-L-iduronidase. This substance, called an enzyme, helps speed the breakdown of long chains of sugar molecules called glycosaminoglycans (formerly called mucopolysaccharides). These molecules are found throughout the body, often in mucus and in fluid around the joints. Without the enzyme, glycosaminoglycans build up and damage organs, including the heart. Symptoms can range from mild to severe.

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Inclusionary and Exclusionary Guidelines (Clinically based guidelines that may support individual consideration and pre-authorization decisions)

N/A

Related Policies

N/A

Medicare Information

This test is covered for Medicare members.

(The above Medicare information is current as of the review date for this policy. However, the coverage issues and policies maintained by the Centers for Medicare & Medicaid Services [CMS, formerly HCFA] are updated and/or revised periodically. Therefore, the most current CMS information may not be contained in this document. For the most current information, the reader should contact an official Medicare source.)

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References

- Muenzer, Joseph, MD, PhD, "The Mucopolysaccharidoses: A Heterogeneous Group of Disorders with Variable Pediatric Presentations," *The Journal of Pediatrics*, May 2004, pp. 27-34.
- Nakanishi, Takeshi, MD, et al., "Association of hyperhomocysteinemia with plasma sulfate and urine sulfate excretion in patients with progressive renal disease," American Journal of Kidney Diseases, Volume 40, Issue 5, November 2002, pp. 909-915.
- Parks, Joan H, et al., "A Single 24-Hour Urine Collection Is Inadequate for the Medical Evaluation of Nephrolithiasis," *The Journal of Urology*, Volume 167, Issue 4, April 2002, pp. 1607-1612.
- Welch, Brian J., MD, "Biochemical and Stone-Risk Profiles with Topiramate Treatment," *American Journal of Kidney Diseases,* Volume 48, Number 4 (October), 2006, pp 555-563.

The articles reviewed in this research include those obtained in an Internet based literature search for relevant medical references through 6/29/09, the date the research was completed.

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Joint BCBSM/BCN Medical Policy History

Policy Effective Date	BCBSM Signature Date	BCN Signature Date	Comments
11/1/09	9/10/09	8/18/09	Joint policy established; policy retired

This is an established policy and no longer subject to routine review.

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BLUE CARE NETWORK BENEFIT COVERAGE
POLICY: URINE SULFATE

I. Short Description:

This laboratory test may be ordered for the purpose of diagnosing certain abnormalities of sulfur metabolism. Sulfates may be measured for the diagnosis of metachromatic leukodystrophy (sulfatide lipidosis), an inherited lipid metabolism disorder that results in the accumulation of metachromatic lipids in the tissues of the central nervous system, leading to paralysis and often death in early adolescence. The specimen is a random or timed urine collection. The testing method used is spectrophotometry.

II. Coverage Determination:

Commercial HMO (includes Self-Funded groups unless otherwise specified)	Covered.
BCNA (Medicare Advantage)	Covered.
BCN65 (Medicare Complementary)	Coinsurance covered if primary Medicare covers the service. Exception: If BCN65 member has an “exact-fill” option, BCN may cover the service even if Medicare does not.
BlueCaid	Covered.

III. Administrative Guidelines:

- The member's contract must be active at the time the service is rendered.
- The service must be authorized by the member's PCP except for Self-Referral Option (SRO) members seeking Tier 2 coverage.
- Services must be performed by a BCN-contracted provider, if available, except for Self-Referral Option (SRO) members seeking Tier 2 coverage.
- Payment is based on BCN payment rules, individual certificate benefits and certificate riders.
- Appropriate copayments will apply. Refer to certificate and applicable riders for detailed information.
- CPT - HCPCS codes are used for descriptive purposes only and are not a guarantee of coverage.

IV. Effective Dates:

JUMP policy effective date: 11/1/09

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