

In addition to the specialty and subspecialty requirements outlined in this standard, all California Children's Services (CCS) Metabolic Special Care Centers (SCC) are required to comply with CCS Chapter 3.37 CCS Provider Core Standards.<sup>1</sup>

**A. Metabolic SCC Overview**

1. Metabolic SCC shall provide care and treatment for patients with CCS-eligible inborn errors of metabolism.
2. The Metabolic SCC staff shall operate as a distinct team responsible for coordinating all aspects of a CCS client's care, including comprehensive evaluation, treatment, and management of pediatric patients.

The Metabolic SCC shall be staffed by a board-certified clinical geneticist with metabolic training, registered dietician (RD) with metabolic training, CCS paneled genetic counselor (GC), and a social worker (SW). Staffing may also include a nurse practitioner (NP), or registered nurse (RN).

3. The CCS Metabolic SCC shall see patients with known or suspected metabolic disease, including but not limited to patients diagnosed with an inborn error of metabolism identified by the California Newborn Screening (NBS) Program including, but not limited to:
  - a. Phenylketonuria (PKU),
  - b. Maple syrup urine disease (MSUD),
  - c. Methylmalonic aciduria (MMA),
  - d. Fatty acid oxidation defects,
  - e. Glycogen storage disorders,
  - f. Lysosomal storage disorders (LSD),
  - g. Mucopolysaccharidosis (MPS), and
  - h. Urea cycle disorders (UCD).
4. The Metabolic SCC shall request appropriate laboratory studies including:
  - a. Diagnostic biochemical studies,
  - b. Routine nutritional labs,
  - c. Molecular testing and imaging studies such as abdominal ultrasounds, echocardiograms, and magnetic resonance imaging (MRI), and

- d. Additionally, the Metabolic SCC may request targeted molecular testing of parents for variants identified in the child.

**B. CCS Program Requirements**

In addition to adhering to the Core SCC Standards Section B, the Metabolic SCC shall:

1. Be affiliated with a tertiary hospital that is able to care for children with inborn errors of metabolism by providing appropriate dietary and therapeutic support during any hospitalization.<sup>2</sup>
2. Be contracted with the NBS Program to accept referrals of newborn screening-positive infants.<sup>3</sup>
3. Satellite centers may be based at a community hospital. Satellite centers must have a CCS-paneled geneticist, SW, nurse, on staff, and be affiliated with a tertiary-hospital-based SCC.

**C. Metabolic SCC Core Team Members and Specialty Consultants**

1. There shall be a core team that meets regularly to evaluate patients, to initiate or modify care plans, and to perform other functions as needed to provide on-going, multidisciplinary care. If possible, the personnel on the core team should be consistently/permanently assigned to the team. The core team and members responsibilities shall meet the criteria described in the CCS Core Standards, Section C.3., and shall consist of the following:<sup>4</sup>
  - a. Medical Director – Board certified in Clinical Genetics and Clinical Biochemical Genetics or Medical Biochemical Genetics through the American Board of Medical Genetics and Genomics.
  - b. SCC Coordinator – this may be a physician, nurse, GC, or SW.
  - c. CCS paneled RN.
  - d. CCS paneled SW – Licensed in California and experienced in working with children and families. Provide social service support to patients and their families.
  - e. GC – Board certified by the American Board of Genetic Counseling and licensed in the state of California with at least two years' experience providing care for patients with inborn errors of metabolism.

- f. CCS paneled RD – Board certified with at least two years of clinical experience and advanced training in inborn errors of metabolism.
2. Other pediatric subspecialists available to the Metabolic SCC – A Metabolic SCC shall have an available referral network should including pediatric gastroenterology and hepatology, pediatric pulmonology, child neurology, physical medicine and rehabilitation (including occupational therapy, physical therapy, and speech therapy), pediatric nephrology, pediatric surgery, pediatric cardiology, pediatric orthopedic surgery, and pediatric endocrinology.
3. The Metabolic SCC shall maintain allied health personnel that are regularly available for consultation, counseling and/or treatment. These allied health personnel shall be CCS-paneled as applicable, and shall include:
  - a. Occupational therapy,
  - b. Physical therapy, and
  - c. Speech therapy.

**D. Metabolic SCC Facilities and Equipment**

Facilities and equipment shall meet the standards described in CCS SCC Core Standards, Section D.

**E. Metabolic SCC Patient Care**

In addition to adhering to the patient care standards in the Core SCC Standards Section E, the Metabolic SCC shall:

1. Provide and recommend standard of care treatment for patients with inborn errors of metabolism including, but not limited to:
  - a. Medications,
  - b. Medical formulas,
  - c. Medical foods, and
  - d. Medically necessary supplements including investigational treatment when appropriate.
2. Refer for medical specialty evaluations as indicated by the diagnosis or clinical findings.

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Special Care Center

3. Coordinate with community resources, such as the National Organization for Rare Diseases, and disease specific support groups including the PKU alliance and the United Mitochondrial Foundation to support patients and families.
4. Inform patients with Genetically Handicapped Persons Program (GHPP)-eligible metabolic conditions about the GHPP program before the patient's 21<sup>st</sup> birthday.

#### **F. Metabolic SCC Quality Assurance (QA) and Quality Improvement (QI)**

In addition to adhering to the quality requirements described in the Core SCC Standards Section F, the Metabolic SCC shall participate in the NBS Program. This includes attending in person and virtual meetings, accepting referrals for the area service center, and providing long-term/follow up data to the NBS Program.

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<sup>1</sup> California Children's Services Special Care Center Core Standards

<https://www.dhcs.ca.gov/services/ccs/Documents/Chapter-3.37-CSS.Core.Standards.pdf>

<sup>2</sup> California Children's Services Provider Standards

<https://www.dhcs.ca.gov/services/ccs/Pages/ProviderStandards.aspx>

<sup>3</sup> California Department of Public Health Newborn Screening Program

<https://www.cdph.ca.gov/Programs/CFH/DGDS/Pages/nbs/default.aspx>

<sup>4</sup> California Children's Services Special Care Center Core Standards

<https://www.dhcs.ca.gov/services/ccs/Documents/Chapter-3.37-CSS.Core.Standards.pdf>