CDCC Program Requirements for Care & Services

Clinical and Sub-Specialty Services

Certified Duchenne Care Centers employee teams of sub-specialists offering comprehensive sub-specialty care and clinical services to people and families living with Duchenne. The each sub-specialist will be expected to provide care according to the CDCCP Standards.

Centers must include access to the following providers at each visit (at the same facility, during the same multi-day or single-day visit):

- Center director
- Clinic coordination, preferably by a nurse and/or nurse practitioner
- Neurologist and/or physical medicine and rehabilitation with training and/or expertise in neuromuscular care
- Cardiology
- Pulmonary
- Genetic counseling (available to see families within 6 months of diagnosis)
- Physical therapy (PT)
- Social work familiar with the diagnosis of Duchenne and the needs of families/patients, available in person or by phone as needed

The following sub-specialty and clinical services must be available as needed to see patients (at the same facility, during the same multi-day or single-day visit) when appropriate:

- Gastroenterology/Urology
- Orthopedics
- Endocrinology
- Palliative care
- Surgery
- Durable medical equipment (DME)
- Nutrition
- Orthotics
- Occupational therapy (OT)
- Psychology/Neuropsychology/Counseling/Developmental pediatrics/Neurodevelopmental Disabilities
- Speech therapy
- Wheelchair specialist

Provision of Care

Care will be provided according to the standards as identified by the Duchenne muscular dystrophy, Care Consideration Working Group, and specified by the Certified Duchenne Care Center Program. Measures for care in the 5 identified domains include

Coordination and Communication

Measures for the Center:

- Designated clinic coordinator (preferably a RN or CNP)
- If patients over 21yo are seen, relationships have been formed with adult sub-specialty providers

Measures for the Patients:

- A copy of the visit note/summary is sent to the primary care provider
- Disease specific information/resources available/sent to primary care providers
- Patients/families receive a copy of the note/summary sent to their primary care provider, or will have access to an electronic copy of their note/summary

Pulmonary

- Patients are measured for height/length by standing (when appropriate) and ulnar length at each pulmonary visit.
- Patients >6yo, for whom it is appropriate, receive standard spirometery FVC, FEV1, FEV1/FVC ratio, FEF 25-75) once while ambulatory and once/twice annually after loss of ambulation by a certified respiratory therapist at a reliable institution/lab.
- Cough peak flow is measured in patients >6yo for whom it is appropriate, at least annually and more often as needed.
- %O2 Sat measured at least annually and more often as needed.
- End tidal CO2, or its equivalent (i.e., blood gasses) is measured at each visit for patients > 6 years for whom it is appropriate
- Capnography is checked at least annually in patients with oxygen saturations <95% and more often as needed
- Patients will be questioned regarding symptoms of sleep related hypoventilation at each visit.
- Sleep studies are encouraged when FVC is <50% predicted; overnight pulse oxymetry may be used if a sleep study is not available.
- Cough assist will be offered when the cough peak flow is <270 lpm, or otherwise indicated.
- Cough assist use is encouraged and practiced during respiratory illnesses while ambulatory, and daily and as needed during respiratory illnesses when non-ambulatory.
- Bi-PAP is encouraged for the management of sleep related hypoventilation when FVC <30% and/or with evidence of sleep related hypoventilation on polysomnogram.
- Daytime intermittent ventilation will be encouraged when exhaled PCO2 >45 mmHg or when Hgb sat <92% while awake.

Cardiology

- Every patient has a clinical visit with a cardiologist at diagnosis or before age 6, then at least bi-annually until age 10, and once/twice annually thereafter.
- Every patient receives an EKG at diagnosis or before age 6, at least bi-annually until age 10, and once/twice annually thereafter, or at the discretion of the clinician.
- Every patient receives cardiac imaging at diagnosis or before age 6, at least bi-annually until age 10, and once/twice annually thereafter.
- Cardiac medications are offered with evidence of decreasing cardiac function of by echocardiogram or MRI, with the onset of heart failure (SF< 28%, EF <55%) and/or with evidence of cardiac fibrosis by MRI, or sooner at the discretion of the cardiac provider.

Neuromuscular Care (Neurology/PM&R)

- Patients receive a baseline x-ray with any clinical evidence of scoliosis while ambulatory, or within 1 year of loss of ambulation, regardless of the presence or absence of signs of scoliosis.
- Scoliosis evaluation is performed annually when the curve is <15-20 degrees and every 6 months when the curve(s) >20 degrees during skeletal growth, and after growth if clinically indicated.
- Patients have longitudinal strength testing as part of clinical care at least annually.
- Patients have a functional assessment as part of clinical care at least annually.
- Night splints are offered at first evidence of plantar flexion contracture and encouraged throughout the life span as indicated in order to delay/prevent deformity.
- Patients have OT/PT evaluations every 4-6 months to prevent contractures.
- Patients see an OT/PT/PM&R provider at least annually for evaluation and recommendations.
- The use of corticosteroids, and their side effects will be discussed with every person with Duchenne, or their parent, over age 3.
- Every patient, for whom it is appropriate, will be prescribed a corticosteroid by age 6yo or before.
- Continued use of corticosteroids throughout the lifespan is encouraged.
- Neurodevelopmental/psychosocial topics (risk for learning disability, behavioral disorder (ADD/ADHD, autism, OCD), need for IEP or 504 Plan (for cognitive or physical indications), need for neurocognitive evaluation, risk for social isolation of the person living with Duchenne and/or parents) will be discussed at least once with parents and followed up on appropriately.
- Informal assessments for emotional adjustment/coping, speech/language, signs of autism, signs of behavioral disorder (ADD, ADHD, OCD), signs of emotional disorder (anxiety, depression) and social isolation will be performed at each visit.

Genetic/Genetic Counseling

- Each patient is encouraged to have genetic testing.
- Genetic counseling is available at each clinic, and will include genetic risk, carrier testing and pre-implantation genetics (as appropriate)

Endocrine and Bone Health

- Patients are weighed at each visit, standing or sitting; pre-recorded wheelchair weights are not recommended.
- Height/length is measured at each visit during growth, and annually thereafter, either by standing and/or by ulnar length.
- Pubertal development by Tanner staging is annually performed by either primary care and shared by parent/patient report, or performed by endocrinology
- Patients taking corticosteroids receive bone density testing within 1 year of beginning corticosteroids and repeated every 1-2 years.
- Patients not taking corticosteroids receive bone density testing within a year of loss of ambulation and repeated every 1-2 years.
- Patients taking steroids have 25-OH vitamin D checked just prior to starting corticosteroids and repeated annually.
- Patients taking corticosteroids are screened annually for signs/symptoms of DM type II and I.
- Patients taking corticosteroids with signs/symptoms of DM type I and/or II have an oral glucose tolerance test (OGTT) for definitive diagnosis.
- Patients are referred to endocrinology for any issues regarding height/growth/pubertal delay and bone health.