Caring for Persons with Muscular Dystrophy

Muscular dystrophies are characterized by progressive muscle weakness and atrophy. Duchenne and Becker types are related conditions, primarily affect skeletal and cardiac muscle. Occur almost exclusively in males. Together affect 1:3500 - 5000 newborn males as X-linked recessive. Different mutations in the DMD gene alter instructions for the protein dystrophin located primarily in skeletal and cardiac muscle. Dystrophin helps stabilize and protect muscle fibers and plays a role in chemical signaling within cells.

System Information Cognition I Qaverage ~ 1 SD below norm, limits esp. in working memory, increased dyskeria, dyscaiculia, dysgraphia w/ 20% moderate + 20% serious reading problem. Co-morbidities 23% sustism, 35% ADHD, and OCD. Neurology Progressing muscle weakness, affects hip/legs early. New treatments include gene therapy, exon skipping treatments, novel corticosteroids and histone deacetylase enzyme inhibitor. Pulmonary Respiratory failure with decline in function ~ 4-6.9%/ yr from age 9, some improve/delay w/ corticosteroids. Night arousals, day sleepiness from nocturnal desaturation and hypercapnia. Progressive hypoventiation/ atelectasis/ reduced cough assoc w/, lung infections. PCF > 270 l/min = effective cough. If cushingoid, think about OSA. Poor swallow causes aspiration. Need for ventitatory support increases as FVC < 50%, particularly FVC < 30%. ENT Sialorrhea Cardion-vascular Cardion-vascular Cardion-vascular Progressive GFR decreases, often w/ underhydration and hyperension. 10% renal stones. Small capacity, hyperreflexic bladder, detrusor sphincter dyssynergia. Heme Assess risk for DVT/PE GI Steroids induce overweight. Adult - higher rates of cachexia w/ dysphagia. High rates constipation, Gi dysmotility, GER. Endocrine Responsible of the cardion, pubertal delay d/t hypogonadism, osteoporosis, adrenal insufficiency, frable skin, Gi perforation, pubertal delay d/t hypogonadism, osteoporosis, adrenal insufficiency, frable skin, Gi perforation, pubertal delay d/t hypogonadism, osteoporosis, adrenal insufficiency, frable skin, Gi perforation, pubertal delay d/t hypogonadism, osteoporosis, adrenal insufficiency, frable skin, Gi perforation, pubertal delay d/t hypogonadism, osteoporosis, adrenal insufficiency, fare embolism d/t osteoporosis, adrenal insufficiency, fare embolism d/t osteoporosis, adrenal insufficiency, fare embolism d/t osteoporosis, adrenal insufficiency, fare embolism d/to steeporosis, adrenal insufficiency, fare embolism d/to steeporosis, adrenal insufficiency, far			
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Determine Out in Brown Detail Adult Manth Ctan Material / ANICAL C	Mental health	 17–29% depression and/or anxiety, increases as adult, social isolation, grief. 	 Use mental health, palliative care and advance care planning.

Reference: Quinlivana R et al. Adult North Star Network (ANSN): Consensus Guideline of Standard Care for Adults with DMD. *J Neuromusc Dis*, 2021.

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