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Prader-Willi Syndrome (PWS) is a rare genetic disorder with a multitude of problems, often attributed to hypothalamic dysfunction. A child with PWS has a genetic predisposition to develop obesity due to appetite dysregulation, hyperphagia and excess calorie intake on the one hand; hypotonia, decreased muscle mass and decreased ability to spend the calories on the other hand. Although, there is no cure for PWS, lives of children with PWS can be significantly improved with specialist multi-disciplinary care and obesity should not be considered or viewed as an inevitable end point. Optimal management of growth and body composition are discussed, including growth hormone therapy; active calorie restriction and dietary monitoring; improving physical activities; supporting families to overcome challenging behaviours and overall helping PWS children lead healthy and fulfilling lives.