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Objective evaluation of muscle strength in infants with hypotonia and muscle weakness

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ABSTRACT

The clinical evaluation of an infant with motor delay, muscle weakness, and/or hypotonia would improve considerably if muscle strength could be measured objectively and normal reference values were available. The authors developed a method to measure muscle strength in infants and tested 81 typically developing infants, 6–36 months of age, and 17 infants with Prader–Willi Syndrome (PWS) aged 24 months. The inter-rater reliability of the measurement method was good (ICC = .84) and the convergent validity was confirmed by high Pearson's correlations between muscle strength, age, height, and weight (r = .79–.85). A multiple linear regression model was developed to predict muscle strength based on age, height, and weight, explaining 73% of the variance in muscle strength. In infants with PWS, muscle strength was significantly decreased. Pearson's correlations showed that infants with PWS in which muscle strength was more severely affected also had a larger motor developmental delay (r = .75).

1. Introduction

Making a diagnosis in a hypotonic infant within the first year of life is challenging for pediatricians and pediatric physiotherapists. Despite advances in genetic technology and brain imaging techniques, the value of the clinical assessment cannot be overemphasized and ought to be the first step toward a diagnosis (Birdi, Prasad, Prasad, Chodirker, & Chudley, 2005; Harris, 2008; Paro-Panjan & Neubauer, 2004; Prasad & Prasad, 2011). The clinical assessment of 'floppy' infants focuses

Abbreviations: BSID-II, Bayley Scales of Infant Development 2nd edition; HHD, hand-held dynamometry; ICC, Intraclass correlation coefficient; IMS, infant muscle strength; IMS-meter, infant muscle strength meter; MMT, manual muscle testing; PWSMT, Prader Willi syndrome.

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on differentiating between hypotonia of central (60–80%) or peripheral (15–30%) origin (Harris, 2008; Prasad & Prasad, 2011). Regardless of the anatomical substrata and etiology, hypotonic infants show diminished resistance in the muscles to passive movement, a frog-like posture, excessive joint mobility, inability to maintain normal posture against gravity, and motor developmental delay (Prasad & Prasad, 2011). If, despite these features, muscle strength is relatively normal then a central origin is most likely, whereas profound muscle weakness indicates a peripheral origin (Bodensteiner, 2008; Harris, 2008; Paro-Panjan & Neubauer, 2004; Prasad & Prasad, 2011). Evaluation of muscle strength in infants is difficult since no objective measurement methods are available. Clinicians observe the antigravity movements of the infant and test muscle strength manually using several maneuvers, such as the so-called 'pulling to sit', 'scarf sign', 'shoulder suspension', and 'ventral suspension' (Bodensteiner, 2008; Martin, Kaltenmark, Lewallen, Smith, & Yoshida, 2007). However, on the basis of these observations it is still difficult to determine whether the infant suffers from both hypotonia and muscle weakness or from hypotonia only (Bodensteiner, 2008). Quantifying muscle strength in infants would enable us to determine to what extent muscle strength is diminished in relation to typically developing infants, and to evaluate effects of treatment on muscle strength.

Techniques to assess muscle strength in children and adults are well established and have been validated. Examples are manual muscle testing (MMT), hand-held dynamometer (HHD) and isokinetic dynamometry. MMT is used in accordance with the Medical Research Council scale, in which strength is classified on a 5-point scale (Mendell & Florence, 1990). When performed by very experienced assessors, the intra- and inter-rater reliability of MMT are satisfactory (Escolar et al., 2001; Florence et al., 1992). Unfortunately MMT has a limited sensitivity and specificity for detecting muscle weakness, particularly in patients with mild muscle weakness (Bohannon, 2005). HHD has been validated (Li et al., 2006; Stark, Walker, Phillips, Fejer, & Beck, 2011) and its intra- and inter-rater reliability are very satisfactory for children (Escolar et al., 2001; Macfarlane, Larson, & Stiller, 2008; Van den Beld, Van der Sanden, Sengers, Verbeek, & Gabreels, 2006), even children as young as 30 months of age (Gajdosik, 2005; Rose, Burns, Ryan, Ouvrier, & North, 2008). Isokinetic dynamometry is considered to be the golden standard. It is not used regularly for children, as the complex equipment used for adults requires extensive adaptations to fit to the various anthropometric characteristics of children (Jones & Stratton, 2000). Unfortunately, none of these methods are suitable, as infants are not able to generate maximum muscle strength in a muscle group on demand (Gajdosik, 2005; Rose et al., 2008; Van den Beld et al., 2006). Therefore, we developed a new measurement method to quantify muscle strength in infants and toddlers, the so-called "Infant Muscle Strength meter" (IMS-meter). In infants aged 6 months and older, it is natural to pull a desirable object. Based on this behavior, we developed a pulling task to evoke a maximal pulling action and we then determined whether maximum muscle strength during such a spontaneous reaction in typically developing infants and toddlers between 6 and 36 months of age can be measured reliably. Because another measurement method to measure muscle strength in infants is lacking, it is not possible to compare the IMS-meter to another muscle strength test. Therefore, the convergent validity was evaluated by testing its correlation with age, height, and weight, because muscle strength is highly related to these factors (Beenakker, van der Hoeven, Fock, & Maurits, 2001; Eek, Kroksmark, & Beckung, 2006; Rose et al., 2008). A model was developed based on sampled data in a convenient cohort of typically developing infants. With this model a prediction for typical muscle strength can be made, and this prediction can be compared to muscle strength in hypotonic infants. As an example to show how the IMS-meter can reveal understanding into what extent muscle strength is affected in hypotonic infants, we tested 24 months old infants with Prader–Willi syndrome (PWS), in which hypotonia, muscle weakness, and seriously delayed motor development are characteristic (Cassidy & Driscoll, 2009; Reus, van Vlimmeren, Bart, Otten, & Nijhuis-van der Sanden, 2012).

2. Methods

2.1. Participants

Typically developing infants, divided in seven age groups of 6, 9, 12, 18, 24, 30, and 36 months (age range ±21 days), were recruited in 2010 through an advertisement in a regional newspaper and an information leaflet in Bernhoven Hospital (general hospital) in Veghel, The Netherlands. Information about the child's health status was obtained from the parents during a telephone contact. If the infant was born at term, healthy, and typically developing, an appointment was scheduled. All parents gave written informed consent and the study was approved by the Medical Ethical Committee. All participating infants with PWS visited the department of pediatric physical therapy at the Radboud University Nijmegen Medical Center regularly. For this study we used muscle strength data measured with the IMS-meter at the age of 24 months. All parents give written informed consent and the study was approved by the Medical Ethical Committee.

2.2. Measurements

For all children, height in centimeters and weight in grams were measured using a calibrated length meter and weight scale. Motor performance was assessed with the Motor Scale of the Bayley Scales of Infant Development, 2nd edition (BSID-II). This is a norm-referenced test to assess gross and fine motor skills between the age of 1 and 42 months (Bayley, 1993; Provost, Crowe, & McClain, 2000). In the typically developing infants, from the raw scores a psychomotor developmental index (PDI) was obtained to verify if motor development was within the normal range (PDI mean (100) ± 1 SD (15)). Because of severe motor developmental delay in infants with PWS PDI-scores are all below −2 SD (Festen et al., 2008), therefore, we