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Research review

A review of chemosensory perceptions, food preferences and food-related behaviours in subjects with Prader–Willi Syndrome

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ABSTRACT

Hyperphagia and obsessive preoccupation with food are hallmark characteristics of Prader–Willi Syndrome (PWS). Although hyperphagia in PWS is linked to hypothalamic dysfunction, the underlying mechanisms behind this problem are poorly understood. Moreover, our understanding of how chemosensory perceptions and food choice/preferences relate to hyperphagia in individuals with PWS is very limited. This narrative review synthesizes studies that assessed chemosensory perceptions, food choices and food-related behaviours in PWS individuals and highlights knowledge gaps in research for further exploration. Twenty seven publications from relevant databases met inclusion criteria and were organized thematically by study technique in the review. Results suggested that PWS individuals have consistent preferences for sweet tastes and in most studies have exhibited a preference for calorie-dense foods over lower calorie foods. No firm conclusions were drawn concerning the chemosensory perceptions of PWS individuals and their influence on food preferences or choices; chemosensation among PWS individuals is an understudied topic. Current evidence suggests that eating behaviour in PWS is a complex phenomenon that involves a dysfunctional satiation and not excessive hunger. Food preferences, choices, and related behaviours and the impact of these on obesity management in those with PWS remain poorly understood and require further study using validated tools and methodologies.

Contents

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1. Introduction

Hyperphagia, an extreme form of overeating leading to life-threatening obesity, is observed in many populations including but not limited to those with genetic disorders (Amor, 2002; Sherafat-Kazemzadeh et al., 2013; Welham et al., 2015), dementia (Keene & Hope, 1998; Keene, Hope, Rogers, & Elliman, 1998; Smith, Vigen, Evans, Fleming, & Bohac, 1998), hypothalamic lesions (Roth, Wilken, Hanefeld, Schroter, & Leonhardt, 1998; Shinoda et al., 1993) and otherwise healthy adults (Heymsfield et al., 2014). Hyperphagia is difficult to treat and remains one of the most detrimental features of Prader–Willi Syndrome (PWS).

PWS is a common form of syndromic obesity that affects ~1: 15,000 births with equal gender and ethnic distribution (Benarroch et al., 2012; Dykens, Maxwell, Pantino, Kossler, & Roof, 2007). PWS is identified using genetic testing and/or clinical diagnostic criteria (McCandless & Cassidy, 2006) and is caused by a lack of paternal gene expression on the long arm of chromosome 15 (Brondum-Nielsen, 1997). Approximately 70% of PWS cases are the result of the absence of genetic information from the paternal copy of chromosome 15 and 20–30% of cases are due to maternal uniparental disomy (UPD). The remaining 1–5% of cases are secondary to rare mutations in the PWS imprinting locus (Chen, Visootsak, Dills, & Graham, 2007; Whittington et al., 2004).

PWS is characterized by a variety of features including physical, physiological, cognitive and behavioural abnormalities (Chen et al., 2007; McCandless & Cassidy, 2006; Pignatti et al., 2013). The eating behaviour of individuals with PWS is characterized by two distinct nutritional phases (Chen et al., 2007; Rankin & Mattes, 1996). During the first phase in early infancy, PWS individuals present with severe hypotonia, delayed motor development, poor sucking reflexes and disinterest in food; these lead to failure to thrive (Chen et al., 2007; McCandless & Cassidy, 2006). The second phase, between 1 and 6 years of age, is characterized by insatiable appetite, progressive weight gain, obsessive food-seeking, and interest in inappropriate food (Chen et al., 2007; Rankin & Mattes, 1996). More recent studies have suggested a more complex progression of the syndrome, with seven identified nutritional phases, beginning with growth restriction in utero. After birth, the phases progress through hypotonia in infancy (up to a median age of 9 months; with or without failure to thrive), normal growth (9–24 months of age), and a phase of weight gain with no change in appetite (2–4.5 years) and further weight gain associated with increased interest in food (4.5–8 years). A subsequent phase from 5 to 13 years of age involves food seeking and lack of satiety, which some suggest may progress to a satiable appetite (feeling of fullness) in adulthood (Cassidy, Schwartz, Miller, & Driscoll, 2012; Miller et al., 2011).

Hyperphagia in PWS, which is linked to hypothalamic dysfunction, causes a lack of satiety after eating and can lead to choking, gastric perforations, life-threatening obesity and obesity-related comorbidities (Holland et al., 1993; Zipf & Berntson, 1987). Since there is no effective medication to ameliorate hyperphagia (Benarroch et al., 2012), its management remains mainly behavioural and preventive (Whitman & Thompson, 2006). Much has been learned about the unique medical, developmental and behavioural issues of individuals with PWS (McAllister, Whittington, & Holland, 2011; Rice & Einfeld, 2015; Whittington & Holland, 2010) and the role of appetite-regulating hormones (Scerif, Goldstone, & Korbonits, 2011; Tauber et al. 2014) and appetite stimulation (Hinton, Isles, Williams, & Parkinson, 2010) in PWS hyperphagia. However, our understanding of hyperphagia and its relationship to chemosensory perceptions, food preferences and food-related behaviours among individuals with PWS is very limited. Thus, the objective of this narrative review was to describe studies designed to 1) assess chemosensory perceptions, food preferences and food-related behaviours in PWS individuals, 2) evaluate the state of knowledge in these areas and 3) highlight knowledge gaps in research for further study.

2. Methods

2.1. Search strategy

Relevant literature was identified from the year of database initiation until March 31, 2015 using the following electronic databases: MEDLINE (OvidSP), EMBASE (Excerpta Medica Database, OvidSP), CAB (Commonwealth Agricultural Bureaux) and FSTA (Food Science and Technology Abstracts). Three different searches were carried out, with no limits or filters. The first search included the following MeSH and free-text terms: “Prader–Willi Syndrome or PWS” and “food or food preference* or food deprivation* or food habit* or food quality or food supply or food choice* or feeding or motivation or food motivation”. The second search included the following MeSH and free-text terms: “Prader–Willi Syndrome or PWS” and “taste or taste threshold or taste perception or taste disorder or taste buds”. The last search included: “Prader–Willi Syndrome or PWS” and “eating or eating disorders or hunger or satiation”. The search terms were adapted for each database. Additionally, a hand search through reference lists of relevant articles was performed. Publications such as reviews, editorials, letters, abstracts or expert opinions were excluded.

2.2. Data extraction

Resulting articles were evaluated for inclusion, obtaining full text of those identified as meeting criteria. Data extracted from each eligible publication included sample characteristics, a description of the assessment methods and study findings.

3. Results

3.1. Literature search results

The initial search identified 407 unique articles. Of these studies, only 35 publications met inclusion criteria and were extracted for full article review. A further 10 articles were excluded that were either reviews (n = 4), related to the use of food as a reinforcer (n = 4), reports (n = 1) or opinions (n = 1). One article was added from the reference lists of relevant articles and two were added during the review process; 27 articles were included in this review (Fig. 1). For presentation in this review, studies are organized thematically by the techniques used to assess chemosensory perception, food preferences and food-related behaviours.