

A Case Of Prader-Willi Syndrome With Behavioural Disturbances: A Successful Multidisciplinary Management Approach

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ABSTRACT

Prader-Willi Syndrome (PWS) is a genetically determined neurodevelopmental disorder occurring in 1 in 15,000 births. PWS is a rare case in Malaysia and a successful approach to its management has not been well reported here. We present a case of a 13-year-old boy with Prader-Willi Syndrome with prominent behavioural disturbances characterised by temper tantrums, compulsive food intake, stubbornness, stealing and impulsivity further complicated by underlying morbid obesity, poorly controlled type 2 diabetes mellitus, hypertension, dyslipidaemia, obstructive sleep apnoea syndrome and intellectual disability. Multidisciplinary approach involving child and adolescent psychiatrist, occupational therapist, counsellor, family therapist, endocrinologist and dietician has shown to improve the patient's weight, glucose and blood pressure control and most importantly the behavioural disturbances.

KEYWORDS: Prader-Willi Syndrome, adolescent, temper tantrums, behaviour disturbances, intellectual disability

INTRODUCTION

Prader-Willi syndrome is a genetically determined neurodevelopmental disorder caused by the absence of paternal expression of imprinted genes in the critical region at 15q11–13 with an incidence of 1 in every 15,000 births [1]. The main characteristic features include neonatal hypotonia, feeding problems, short stature, hypogonadism, hyperphagia, obesity, characteristic facial appearances and mild to moderate intellectual disability [2].

Prader-Willi syndrome leads to an increased risk for specific comorbid behavioural and psychiatric problems [3]. There is initial extreme hypotonia and failure to thrive in newborns, followed by early childhood preoccupation with food and hyperphagia, developmental and cognitive delay, evidence of relative growth and sex hormone deficiency (short stature and

impaired sexual development) and high ghrelin levels. There are also marked propensity to problem behaviours, such as temper outbursts, repetitive and ritualistic behaviours, mood swings and skin picking [4]. In Malaysia, it is a rare opportunity to see patients with PWS. This case report is written to discuss the journey of recovery for an adolescent with PWS.

CASE PRESENTATION

IR is a 13-year-old Malay boy, the eldest of 3 siblings, with a history of hyperphagia, stubbornness, frequent temper tantrums, emotional lability, stealing, occasional aggressive behaviour and sexual disinhibition presenting to the Child and Adolescent Psychiatry Clinic. He has underlying Prader-Willi Syndrome since birth confirmed by genetic testing. He also has comorbidities of morbid obesity, type 2 diabetes



mellitus, hypertension, dyslipidaemia and obstructive sleep apnoea.

Both his parents have stable jobs and hold significant posts. IR was given a lot of focus and attention since young due to his illness, which required constant medical attention and frequent hospital admissions. There are evidence of inconsistent parenting styles between his parents. While the father is strict, his mother often gives in to IR's demands. The family lives with IR's maternal grandparents who became his sole carers when his parents were at work during the day.

His grandparents struggled in managing his behavioural outbursts and they frequently gave in to his demands especially when it concerns food. IR would hit his grandmother and overpower her if she tried to restrict his food access. His parents were the only ones who were able to monitor and control his dietary intake or motivate him to exercise. This only occurred when they were around.

Driven by his hyperphagia, IR would steal money at home and school to buy food. At home, IR was expected to limit his food intake and choices of food while everyone else (his cousin's who stayed together) were not on any rationing. This lead to IR feeling dissatisfied and subsequently sneak-out food and throw tantrums frequently when his demands were not met. These behaviour problems increased in frequency and intensity as he approached adolescence and subsequently started to interfere with his glucose control.

IR had multiple hospital admissions for blood sugar stabilisation and weight-loss program. He struggled to control his blood sugar levels when treated on outpatient basis as opposed to optimal glucose control when warded. This is evidenced by a stable glucose reading of less than 10mmol/L. A few days post discharge, his blood sugar levels would surge up to more than 20mmols/L. Presence of proteinuria and high blood pressure were also subsequently detected.

IR was in special education class due to his learning disability. However, upon showing improvement in class, IR was consequently enrolled into mainstream class with the help of his teacher. Unfortunately, IR was not able to catch up with the rest of the class also had difficulty fitting in with his

classmates. Apart from stealing money from the other students, he also went in and out of the classroom during lessons as he pleased. He was once caught smoking cigarettes within the school compound, in which he claimed that the cigarette was given to him by a senior student. There were also two separate incidents in which IR displayed sexual disinhibition by touching a female student's chest along the school corridor. He explained that he just felt like touching the female student. There were no further details obtained.

Mental state examination revealed an obese boy with almond shaped eyes, small hands and fingers. He was noted to be shy initially but was able to build rapport later. He was able to relate well to his parents and sat quietly while drawing a picture of a thin boy in school uniform. There was good eye contact with normal activity and energy level. He was only able to understand and respond to single and short questions. He appeared not to be able to understand the reason of why his parents were upset with his eating habits and temper tantrums. Estimated intelligence level is lower than his chronological age.

He was managed by a multidisciplinary team. The child and adolescent psychiatrist are the main care provider. Treatment includes one to one session with the patient on regular basis and also combined sessions with the parents to review the progress. Occupational therapists assisted IR with anger management skills, counsellors provided supportive psychotherapy, endocrinologists and dieticians managed blood sugar controls, weight and other endocrine related issues. All disciplines played an important and vital role ensuring optimal management of the patient as everyone complemented and supported each other's efforts.

Family assessment was carried out to address current issues regarding the patient. Temper tantrum was the most significant issue reported by the parents followed by stealing and inappropriate behaviour at school. Behaviour interventions were recommended. A reward system was implemented in the shape of a star chart followed by increasingly satisfying rewards; i.e. allowed to enjoy his favourite food once a week upon good behaviour management. The good behaviour changes were indicated by the number of stars he received over time which was also related directly in better sugar control.

Family therapy was carried out to address the inconsistency of parenting styles and also standardising the family daily menu. Subsequently, the family food intake was standardised for the whole family which eventually reduced IR's previous dissatisfaction. This also led to a more consistent messages from his parents which made IR less confused and enabling him to abide to the rules much easier. Daily routines for IR and his family members became more structured.

Anger management and social skills were also addressed. This resulted in less tantrums and disruptive behaviour. These interventions were adopted by parents and teachers at school in order to ensure the positive behaviours being reinforced continuously. The treating child psychiatrist received regular feedback from the teachers and parents to ensure that the appropriate management was carried out and also to provide support and assistance.

IR was started on Tablet Quetiapine 25mg daily for the impulsivity and anger issues. Both the pharmacological and non-pharmacological approach led to a pre-meal glucose control below 10mmols/L. He lost 3 kilograms within a month and the blood pressure was maintained within the normal range.

DISCUSSION

In Malaysia, it is a rare opportunity to encounter patients with PWS. There are also limited publications available on PWS locally and when available, the issues discussed comprises more of genetics, endocrine and sleep [5-8]. There are minimal, if any, discussion on behavioural issues in PWS. As mentioned in the paper by Benarroch et al. [9], "The multiple facets of the clinical problems demand a multidisciplinary approach with anticipatory medical and psychiatric care, oriented to enhancing the quality of life of individuals who have Prader-Willi syndrome." There is a high need for multiple expertise in managing such cases and as for this case, it involves child and adolescent psychiatrist, occupational therapist, counsellor, family therapist, endocrinologist and dietician.

This case report suggests that there is a need for multidisciplinary approach for patients with PWS which is further complicated by behavioural issues and multiple medical comorbidities. According to Ho et al.

[10] due to the range of problems and variability of symptom severity across individuals with PWS, management of PWS is age-dependent, multidisciplinary, and utilises a problem-based approach to cater to each individual. Hence, each patient with PWS may need a different approach strategy. They also found that behavioural problem when detected early, treatment becomes most effective as the issues and difficulties usually increases with age. These findings are similar to Dykens et al. [11] in which they found that behavioural symptoms including tantrums, food theft, lying, overeating increases with age in PWS patients and the highest rate of maladaptive behaviour in ages around the 20's were due to stressors such as adjusting to a new environment from school to work, as an example [12].

According to Tunncliffe et al. [13], various setting events increased and reduced the likelihood of temper outbursts. The most common antecedent was a change to routine or expectation. There were marked similarities in the sequence of behaviours and emotions during temper outbursts, with anger rising which was quickly followed by expressions of remorse and distress at the end of the outburst. The sequence of behaviours and emotions within outbursts was similar to that described in temper tantrums in typical development. Cognitive and emotional processes are likely to be important in understanding temper outbursts with implications for an early intervention. Hence, it is important for parents, teachers and caregivers to be aware of any changes in the environment that may precipitate the patient's anger outbursts or temper tantrums.

As for the pharmacological treatment, to date, no consensus on usage of psychotropics in the management of behaviour disturbances and temper tantrums in PWS exists. Mohapatra and Panda [14] reported a reduction in stubbornness, irritability and temper tantrums in their 9 years old patient with PWS patients over 8 weeks duration after prescribing risperidone. More research is needed in this area.

Finally, constant communication between all members that play a role in managing the patient is key in order to avoid any loopholes or any miscommunications as a number of interventions occur simultaneously in the interest of benefitting the patient.

It would be regrettable should all the effort go to waste due to poor communication among treating members. Therefore, in an ideal situation, it is best that a case manager is assigned to overlook the case and update the other team members regularly [15].

CONCLUSION

Multidisciplinary approach is essential in managing patients with PWS especially for patients similar to IR in view of the nature of presentation. Recurrent admissions combined with pharmacological treatment has shown to be very beneficial for our patient as opposed to rendering pharmacological treatment on outpatient basis. This is largely due to the fact that there are multiple psychosocial factors influencing control of the illness that needs to be taken into consideration. This is to ensure optimal control of medical issues with the sole hope that individuals with PWS can live a full and meaningful life with their loved ones.

Conflict of Interest

Authors declare none

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