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Inclusion of chiropractic care in multidisciplinary management of a child with Prader-Willi syndrome: a case report☆

Rebekah A. Wittman DC, DACCP[a,*], Sharon A. Vallone DC, FICCP[b]

[a]*Clinic Doctor, Kentuckiana Children's Center, Kentuckiana Center for Education, Health, and Research, Inc., Louisville, KY 40206*
[b]*Chair of the Board, Kentuckiana Children's Center, Kentuckiana Center for Education, Health, and Research, Inc., Louisville, KY 40206*

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Abstract

Objective: The purpose of this paper is to present a case of a child with Prader-Willi syndrome and the observed improvement in the degree of scoliosis, immune function, and behavior documented during the course of her treatment.

Clinical Features: A 7-year-old girl presented to Kentuckiana Children's Center with a 15° lumbar levoscoliosis and diagnosis of Prader-Willi syndrome.

Intervention and Outcome: The treatment plan consisted of chiropractic adjustments, craniosacral therapy, movement therapy, and nutritional therapy. Over the course of treatment, her muscle strength, tone, and motor activity increased. She improved in coordination of gait and balance. Over the course of 3 years, her scoliosis decreased to 4° to 5°. Improvements in immune function and a reduction in anxiety type behaviors were documented by the parents and doctor of chiropractic over the course of 5 years.

Conclusions: This case report describes the improvements and progression of one female child with Prader-Willi syndrome under chiropractic and multidisciplinary care.

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* Corresponding author. Kentuckiana Children's Center, Kentuckiana Center for Education, Health, and Research, Inc., 1810 Brownsboro Road, Louisville, KY 40206. Tel.: +1 502 893 7227x19; fax: +1 502 368 2308.
E-mail address: drbekah@yahoo.com (R. A. Wittman).

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Introduction

Prader-Willi syndrome (PWS) is a complex genetic disorder involving the lack of expression of chromosome 15q11-q13. These genes are imprinted and silenced on the maternally inherited chromosome; and PWS develops when the paternal alleles are defective, missing, or silenced.[1] The prevalence of PWS is estimated to range from 1 in 8000 to 1 in 25 000 births in the United States.[1-2] Prader-Willi syndrome affects females and males equally and can occur in persons of any race. Although diagnosis can be made by clinical diagnostic criteria, the standard of diagnosis is genetic testing through DNA methylation analysis.[1-3] Primary symptoms and behaviors are listed in Figs 1 and 2.

Treatment of PWS commonly involves a primary care physician and a multidisciplinary team that includes endocrinologists, developmental pediatricians, speech and language therapists, occupational therapists, physical therapists, and ophthalmologists. Early developmental intervention and speech therapy are often needed for guided development. Medication is often the first line of management for many of the behaviors listed in Fig 2. However, these behaviors often do not

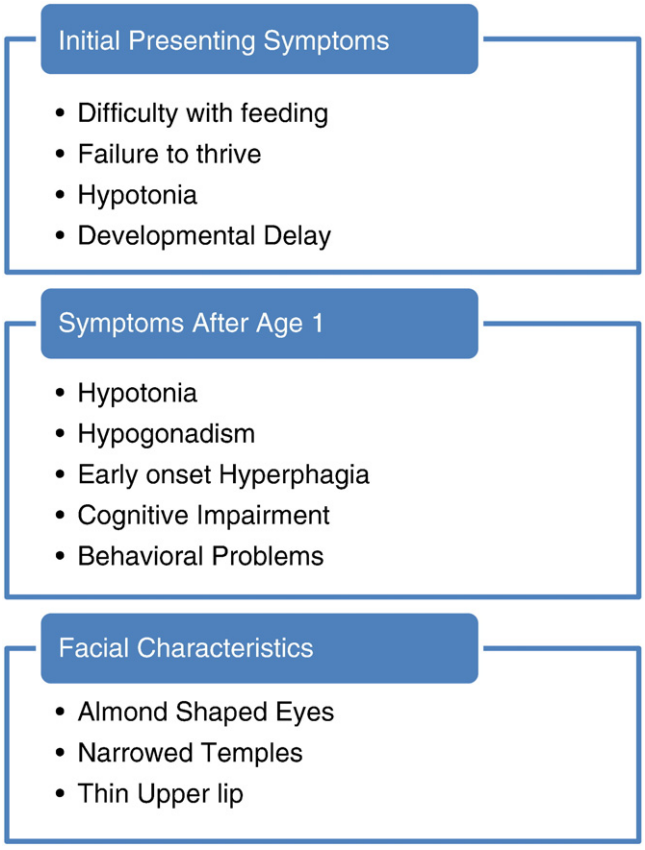

Fig 1. Primary symptoms and presentation of PWS.[1-3]

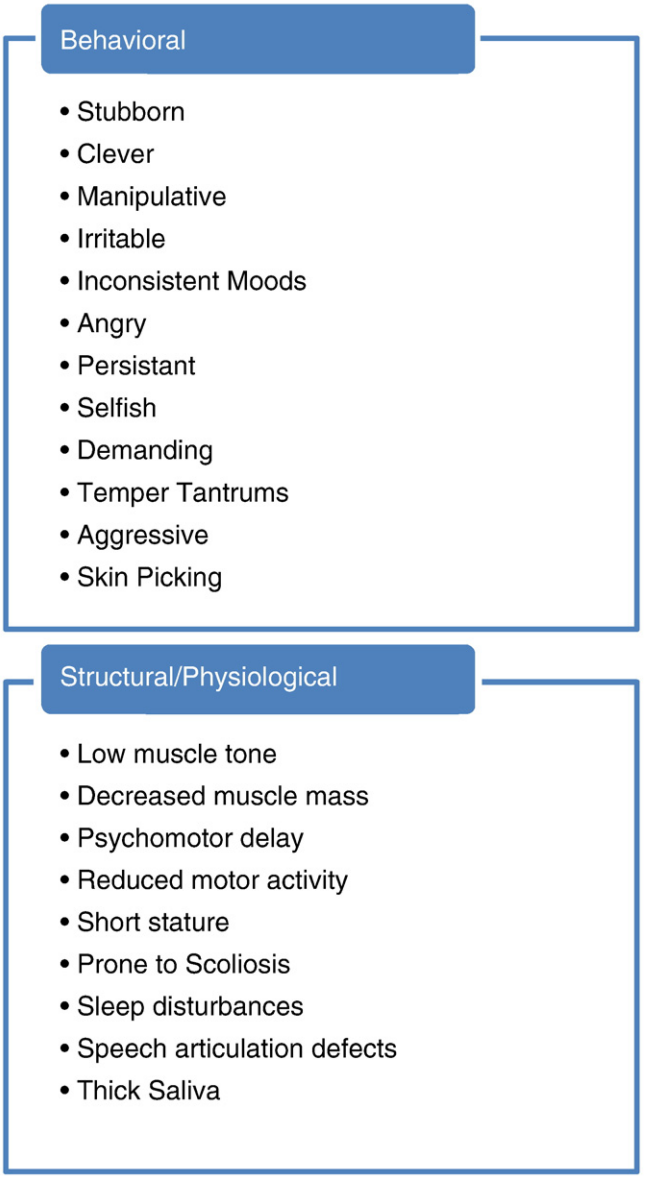

Fig 2. Characterizations of PWS.[2]

respond to psychopharmacologic management; and sometimes, the medications may exacerbate these behaviors.[4] An endocrinologist typically assesses for growth hormone deficiency and monitors growth hormone replacement therapy (GHRT). The GHRT has shown great benefits in body composition and adult height.[1,5-7] Orthopedic doctors typically monitor scoliosis progression.[1-2] Nutrition and dietary intake are strictly monitored to manage hyperphagia and obesity.

Scoliosis is also highly prevalent in PWS.[1,8-12] One study estimated that between 43% and 66% of those with PWS developed scoliosis.[8] The Queen Elizabeth Hospital for Children in London did a retrospective study that demonstrated that 62% of PWS patients had clinical scoliosis.[9] Body composition studies have

shown increased body fat and reduced muscle in PWS, both in infants and adults. These 2 factors are most likely major contributors in the development and prevalence of scoliosis among those with PWS. Unlike idiopathic scoliosis, young children are affected without sex bias.[1] A few studies investigating possible links between GHRT and scoliosis progression have found no direct link and state that any worsening of the scoliosis during this time most likely reflects natural history of scoliosis.[1,8,13] Other studies suggest the clinical need for frequent radiographs of the spine during GHRT to monitor scoliosis progression.[5,8]

The prognosis for adults with PWS depends on the ability to control weight because lack of appetite control leads to excessive eating. Obesity, in addition to the structural and physiologic conditions outlined in Fig 2, has often led to early death due to cardiorespiratory failure.[2,9] However, with early diagnosis, weight control, psychosocial management, GHRT, and use of multidisciplinary physicians, the prognosis is promising.[1]

Chiropractors are sometimes forgotten to be included in the care of children with PWS, despite successful management of spine conditions and case reports of managing idiopathic scoliosis with chiropractic care.[14-16] Chiropractic care of children with PWS is underreported. A literature search using Index to Chiropractic Literature, PubMed, and a public search engine (Google) only produced one case report of a child with PWS receiving chiropractic care. In Arcadi's[17] report, nutritional therapy, chiropractic adjustments, and cranial therapy were used to obtain decreased appetite and improved strabismus, muscle tone, and verbal ability. The following case report outlines the successful management of a child with PWS that included chiropractic care in her multidisciplinary treatment approach.

Case report

A 7-year-old girl was brought to Kentuckiana Children's Center in March 2002 for evaluation and treatment of challenges associated with PWS and a 15° lumbar levoscoliosis. This child was diagnosed with PWS at the age of 3 years. Growth hormone administration began at the age of 5 years, and she was currently taking 1.2 mL of somatropin (Genotropin; Pfizer, Inc., New York, NY) and 18 mg of methylphenidate HCl (Concerta; Ortho-McNeil-Janssen, Inc., Mountain View, CA). Her parents were concerned with adequate nutrition and wanted dietary guidance. She was taking a commercial over-the-counter children's multivitamin daily (Flintstones; Bayer Healthcare LLC, Morristown, NJ). Although she was not overweight, her parents worked diligently to control any hyperphagia. This child had global hypotonia as diagnosed by both her pediatrician and physical therapist. She had poor balance and difficulty focusing. Gross and fine motor skills were delayed, and she had difficulty performing daily tasks such as dressing and hygiene. She was able to feed herself and was toilet trained by the age of 3 years. Her mother felt that she had low endurance and tired easily. She was receiving physical therapy, speech therapy, and occupational therapy; and she was enrolled in a mainstream first-grade classroom.

Social skills were delayed, although she sought out adult interaction. She had anxiety and obsessive compulsive tendencies, which included skin picking and hand wringing. She had a history of multiple illnesses. Her mother noted that she had numerous ear infections and upper respiratory tract infections each year that were treated with antibiotic therapy. She had her first set of tympanostomy tubes inserted at the age of 5 years. The mother reported that the child was absent frequently during her kindergarten and first-grade year because of ear infections, upper respiratory tract infections, or other illnesses. She typically had 1 to 2 bowel movements per day, but was frequently constipated.

This child was born at 35 weeks' gestation by emergency cesarean delivery after a long and difficult labor. Her head shape was normal but edematous after birth. Birth weight was 4 lb 15 oz, and she was 18 in long. Apgar scores were 5 and 8. Skin tags were noted around both ears. She was fed a soy-based formula with iron for 12 months. Solid foods were introduced at 9 months. Development was mildly delayed: she sat on her own at 7 months, crawled at 13 months, and walked at 19 months. Eyesight was evaluated at the age of 3 years, and she had glasses for myopia and astigmatism. The mother noted that the child's prescription needed to be strengthened frequently, approximately every 3 to 6 months. A review of medical records from her endocrinologist indicated that growth hormone treatment was started after a growth hormone stimulation test was given at the age of 4 years that indicated growth hormone deficiency. Coenzyme Q10 was also checked at the age of 7 years that indicated normal levels within the low range.

A physical examination was performed that included vital signs, eyes, ears, nose, throat, chest, abdomen, neurologic, spinal, and cranial examinations. Pupils were responsive to light, but the left eye was dilated

greater than the right because of a prescription eye drop used to help correct the lazy eye. Strabismus was present (Figs 1 and 3). Otoscopic examination revealed excess cerumen and tympanostomy tubes in both ears. Lymph nodes were palpable in the cervical chain and left inguinal chain. Abdominal examination revealed distension as noted with increased tympany. She was able to toe walk, but could not maintain her balance when heel walking. She was noted to have rigid pes planus and hyperpronation. Spinal examination revealed chiropractic subluxations (eg, segmental biomechanical dysfunctions) in the cervical, thoracic, and lumbar spine. Postural examination revealed high right iliac crest and short left leg compared with the opposite side. Radiographs, which were taken at the age of 7 years at a local children's hospital, revealed a 15° levoscoliosis of the lumbar spine with the fulcrum at L1.

At Kentuckiana Children's Center, the treatment plan was devised and coordinated by the staff chiropractor and implemented by the chiropractor and one or more adjunctive therapists. The child's initial treatment plan was implemented in May 2002 and included chiropractic treatments twice weekly for 4 weeks and a diet consultation with the staff nutritionist. The chiropractic technique used included modified activator technique. The nutritionist suggested a trial of removing gluten-and casein-containing foods, while continuing the patient's current low-calorie and low-fat diet.

A trace mineral analysis (Doctor's Data, St Charles, IL) of her hair was ordered and completed in May 2002, just after starting her treatment. Results revealed high levels of uranium and tin and low levels of potassium and manganese. It was recommended that she take coenzyme Q10 at 90 mg/d and 1 oz of a liquid nutrient complex per day to raise her levels of potassium and manganese and to serve as a whole food multivitamin in place of the commercial product she was already taking.

At the end of 4 weeks, the patient progressed to a once weekly schedule of chiropractic and cranial therapy. Movement therapy was added twice monthly in August 2002. Movement therapy goals were to build strength, improve coordination, slow movements, and help increase healthier sleep patterns. The therapist noted that the child had difficulty extending her limbs and that she moved about in a hurried manner. The child was easily distracted and needed constant prompting and redirection. It was also noted by the treatment team that the child was very excitable and did not do well with changes to the treatment schedule.

Fig 3. Presentation of strabismus.

In June 2003, over 1 year from the child's initial examination at Kentuckiana, the treatment planning team recommended that a weekly intensive treatment including chiropractic, cranial work, movement therapy, and art therapy would possibly help to further control anxiety, improve sleep, and support increased focus, while making the schedule more efficient for her parents as well. She was measured every visit to monitor her height and weight. It was also recommended that she begin taking 3 capsules of cod liver oil for the benefits of omega-3 fatty acids in immune function, neurodevelopment, and brain function.[18] Coenzyme Q10 was increased to 60 mg 3 times daily. Dairy products were added back into her diet, but with the use of digestive enzymes with each meal to further aid digestion. By the end of the summer, the child's orthopedic doctor determined that her scoliosis was decreasing and that she no longer needed the rigid shoe orthotics cast by her physical therapist. Through Kentuckiana, she was then cast and placed in flexible leather orthotics (Foot Levelers, Inc, Roanoke, VA) to help maintain pelvic stabilization.

Art therapy was discontinued during the summer of 2004 because of scheduling conflicts with the art therapist. Chiropractic, cranial, and movement therapy was continued on a weekly basis. Dimethylglycine (Kirkman Labs, Lake Oswego, OR) was added at 3 capsules per day for its role in supporting methylation and immune function. Radiographs were ordered in November 2004 to monitor her scoliosis.

Starting in January 2005, she progressed to a twice-monthly treatment plan. She was evaluated by the

clinic's occupational therapist that same month and began working with him on dynamic balance, core stabilization, and endurance during movement sessions. He resumed working with handwriting skills and added breathing exercises to strengthen core muscles and improve sustained breath until May 2007, when occupational therapy was discontinued at Kentuckiana. The child continued to be assessed every other week for both chiropractic and craniosacral therapy.

In addition to therapies received at Kentuckiana Children's Center, this child continued to receive services outside of Kentuckiana such as physical, occupational, and speech therapy. She was also under the care of a pediatrician, endocrinologist, and ophthalmologist.

The initial observable results after starting chiropractic care included improvements in the child's behaviors. Her mother noted better cooperation and more independence. By the end of the first 4 weeks, the child was showing more cooperation in her outside therapies of speech, occupational, and physical therapy. She was also less irritable and more flexible with her schedule at school. Her parents noted increased energy and less fatigability.

After craniosacral therapy and movement therapy were added, improvements in hyperactivity were noted. The child was able to slow down and focus better on specific tasks for longer periods of time. The child often became emotional and irritable when on antibiotic therapy, which was prescribed frequently for ear infections. Her mother and father noted that she was much more tolerant when she received chiropractic and cranial work during an active infection.

The weekly intensive treatments that included chiropractic, cranial work, movement, and art therapy started in June 2003. By December 2003, her scoliosis was maintained; and she was less anxious, was more content, had better focus, was sleeping more soundly, and was showing improvement in her balance while walking. The movement therapist reported that she was better able to use isolated body movements and had better balance. The art therapist reported that she was showing improvement in writing individual letters as long as her feet were on the ground while seated. The craniosacral therapist reported that the child was more tolerant of touch to the back of the head and neck and that she showed improved coordination while getting on the table and was able to lie on her back with less assistance from the therapist. Her mother reported that the child was less sick and anxious.

By November 2004, the child's lumbar Cobb angle was reduced to 4° to 5°. By May 2005, the child was maintaining her height and weight at the 75th percentile. She had gained independence in several areas. She was less anxious and less likely to break down due to an interruption or change in the schedule. Her mother stated that the child did not miss any school for the 2004-2005 year because of being sick compared with an average absence of 1 day a week in the 2001-2002 school year. She had only had 1 case of otitis media in 2005. The child continued to make gains in her balance and coordination skills. Her ability to have a continuous breath (sustained exhale) had improved greatly. She continued to struggle with focus, but this had improved to the point where she could stay on task more easily.

By the end of 2007, she maintained the 4° to 5° of lumbar scoliosis. She was more aware of her body's needs and could vocalize that need to her parents. She was comfortable with her therapies and looked forward to her appointments. She was rarely sick, even when the rest of her family was. Although she still demonstrated mild low muscle tone, she was able to coordinate her body to get on and off tables on her own with ease. Focus and anxiety remained challenges, but her mother noted that she was better able to focus and was significantly less anxious while under active care at Kentuckiana. Gait and balance were greatly improved under active care and were used as a marker for her parents.

Discussion

Many of the characterizations consistent with PWS were evident in this child. She displayed anxiety, poor focus, and persistence. She was demanding, stubborn, and prone to temper tantrums. She was obsessive with skin picking, which often led to bacterial infections. She had poor sleep, speech articulation challenges, and thick saliva. She had low muscle tone, decreased muscle mass, psychomotor delay, and reduced motor activity. However, she was not overweight because of her family's diligence in controlling her diet and limiting her access to food.

Although there are some studies that declare that chiropractic care has no positive effect on reducing scoliotic curves,[19] this case report and several other chiropractic case reports note improvements and reduction in the degree of scoliosis.[14-16,20-21] Romano and Negrini[22] conducted a systematic review of manual therapy as a conservative treatment of adolescent scoliosis. Three of the texts described a combination

of manual techniques along with other therapies.[22] This could suggest that the reduction of the scoliosis is often the result of a combination of manipulative-type therapy and physical exercises, rather than using just one type of therapy. The case reports of Morningstar et al[14,20,23] also use manipulation and rehabilitative procedures as a basis for their success in the reduction of the Cobb angle. The rehabilitative exercises used vary from each other, but all use chiropractic adjustments as a key therapy. The combined treatment plan that was designed specific to this child's needs helped to increase her muscle strength and tone, increased motor activity, improved gait and balance, and subsequently helped reduce the degree of her scoliosis.

This child's family also reported improved immune function based on the occurrence of fewer ear and upper respiratory tract infections after starting her care at Kentuckiana. The mother noted a significant reduction in missed school days and less irritability during an active infection. Literature supporting improvements in nonmusculoskeletal conditions after chiropractic care is often based on subjective findings, such as found in this case report.[24] Cohn[25] conducted a review of literature on the connections between the nervous and immune systems. He concluded that there is growing evidence to support that the chiropractic adjustment can help improve the overall health of a person. The hypothesis used to support Cohn's conclusion is the somatovisceral neural reflex.[24,26-29]

The inclusion of chiropractic in the treatment of children with various conditions is becoming more prevalent. An analysis of 650 children under chiropractic care by Ressel and Rudy[30] demonstrates the many reasons children seek chiropractic care. Because of the complex nature of PWS and the length of this patient's care, it is difficult to assess improvements based on one specific therapy or medication. Prader-Willi syndrome only affects 1 in 8000 to 25 000 births in the United States, which may make it unlikely that a child with PWS will walk into a typical chiropractic office. However, if more research is available to specifically outline the benefits of chiropractic care for conditions other than pain relief or traditional musculoskeletal disorders, then it is the authors' opinion that chiropractic would be more widely accepted as a treatment modality for conditions such as PWS.

Limitations

In our search of literature, there were no articles documenting chiropractic care as a treatment option for scoliosis in an individual with PWS. For that reason, the available case reports regarding chiropractic and scoliosis may not be relevant to this specific genetic disorder. In addition, it should be noted that these referenced case reports all include Cobb angles greater than 20° compared with this child's mild angle of 15°. Children with PWS who have a more severe scoliosis may not respond as positively. As this is only one case, the treatment results as described in this case may not necessarily be applicable to others who have PWS.

Conclusion

This case report demonstrates the improvements and progression of one female child with PWS under chiropractic and multidisciplinary care. Case reports, such as this one, are valuable for the advancement of medicine, educating peers, and sharing new treatment options for a disorder or certain diagnosis. It is unlikely that this child's progress was due solely to her care received at Kentuckiana, but rather her success is due to the combination of therapies both inside and out of Kentuckiana. Often it is not just one specific treatment modality or supplement alone that alters the patient's outcome; rather it is a combination of therapies that are more valuable when combined than when used separately. As in the conventional care of those with PWS, the combination of therapies may achieve the highest results. Based upon the findings of this case report, it is possible that chiropractors may be valuable team members that provide supportive services to multidisciplinary teams that manage conditions such as PWS.

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