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Schroth Therapy for Scoliosis in a Patient with Familial Dysautonomia: A Case Study

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ABSTRACT

Purpose: This case presents a successful physical therapy intervention for the treatment of scoliosis and other neuromuscular disorders in familial dysautonomia (FD) for a patient. **Case Description:** The patient is a 10-year-old girl with FD whose juvenile scoliosis started developing around the age of 7. She presents with a main right thoracic curve and small compensatory curves at the cervical and lumbar areas. She also presents with general muscle weakness, impaired gait, and slight balance deficits. The patient treatment plan consists of conventional physical therapy in combination with Schroth therapy for scoliosis and a Providence nighttime brace. **Outcome:** The patient's Cobb angle reduced by 6° in 3 months and has stabilized since then. Her posture has improved and she is able to perform sports activities involving greater endurance and balancing skills. There has been an overall improvement in the patient's quality of life.

Key Words: Cobb angle, juvenile scoliosis, physiotherapeutic scoliosis specific exercises, Riley-Day syndrome

INTRODUCTION

Familial dysautonomia (FD), also known as Riley-Day syndrome, is a rare genetic disorder of the autonomic nervous system that primarily affects Ashkenazi Jewish people of Eastern European heritage.^{1,2} The disorder is characterized by widespread sensory dysfunction and variable autonomic dysfunction caused by incomplete development of sensory and autonomic neurons. Neuromuscular involvement is seen with hypotonia and spinal scoliosis.

Children with FD present with developmental delays, low muscle tone, an inability to respond to pain and temperature, recurrent pneumonia, hypotension, and ataxia. Atonomic "crisis" is common due to stress triggered by infection or emotional distress. Deep tendon reflexes are often absent due to hypotonia. Impaired renal function, cardiovascular instability, and progressive scoliosis (spinal curvature) are common co-morbidities.¹⁻⁵ Scoliosis is found before the age of 13

years in 90% of children with FD.³

Schroth therapy is a 3-dimensional approach developed by Katharina Schroth in 1921 in Germany. Its principles are based on corrections in sagittal, frontal, and transverse planes.⁶ Schroth therapy uses therapeutic exercises to improve scoliotic posture through elongation and realignment of trunk segments, positioning of arms, and use of a specific breathing pattern (rotational angular breathing).^{6,7}

CLINICAL MANAGEMENT

Clinical management of FD involves a multidisciplinary approach including care by a physical therapist, occupational therapist, speech therapist, neurologist, endocrinologist, pulmonologist, cardiologist, orthotist, psychiatrist, and development specialist. Discussion of each discipline and management of all possible co-morbidities is beyond the scope of this article, which is focused on the treatment of scoliosis. Physical therapy management includes physiotherapeutic scoliosis specific exercises (PSSE), such as Schroth therapy for scoliosis, and conventional physical therapy interventions for improving strength, endurance, posture, and balance.^{6,7} Bracing is effective in reducing and/or delaying further progression of the scoliosis,⁴ but must be managed carefully for rashes and skin pressure ulcers due to lessened pain sensation.⁸

EPIDEMIOLOGY AND ETIOLOGY

Familial dysautonomia is caused by a genetic point mutation in the I-k-B kinase complex associated protein it encodes, IKAP(2-4).¹ Familial dysautonomia is found almost exclusively in people of Ashkenazi Jewish descent, where it affects approximately 1 in 3,700 people. Hayek et al⁹ found that the prevalence of spinal deformity in patients with FD who had lived for at least 20 years was 83%. By the age of 10 years, 52% of patients have scoliosis and 21% have kyphosis with or without scoliosis.

CASE DESCRIPTION

Gina (pseudonym) is a 10-year-old Jewish girl in premenarchal age who presents with

mild to moderate symptoms of FD. She has normal intelligence but delayed physical and emotional development. She was born as a dizygous twin and diagnosed with FD at the age of 3 weeks. Her twin and younger twin brothers are genetic carriers of FD. Gina has been getting physical and occupational therapy at home and school since she was 6 weeks old to develop her motor skills. At age 7, she was diagnosed with scoliosis and her curve progression was monitored regularly by her physician and the school-based physical therapist. By age 10, the curve had progressed to 35° (Figure 1) and she was recommended for Schroth therapy for scoliosis with a night brace in addition to rehabilitation services received in the school setting. School services were provided 3 times a week for a total of 90 minutes.

Schroth therapy is a 3-dimensional based exercise program to correct scoliosis. It uses breathing mechanics, muscle activation, and mobilization. Schroth principles of correction⁶ are based on self-elongation from a neutral pelvis, de-rotation of shoulder, pelvis and rib cage towards neutral, rotational angle breathing to open retracted areas, and stabilization to strengthen balance position. Practitioners using the Schroth method can be certified in the technique after completing a formal education program, although a manual on the technique is available online (Schroth website <http://www.schroth-method.com>). The author (HB) is certified in the Schroth method and initiated Schroth therapy for the patient.

CLINICAL EXAMINATION

Gina attends school full-time and is independent in activities of daily living (ADLs). She has a normal IQ and enjoys arts and crafts, but she was unable to participate in any sport activities due to insufficient endurance for community ambulation and outdoor recreation.

She is on the prescription medication Zantac, and she gets nebulization every day with Pulmicort (0.5 mg) and hypertonic saline. She wears an airway clearance vest for 20 minutes daily. She is also taking various supplements including Tocotrienols, green

tea extract, olive leaf extract, folic acid, omega 3, calcium, vitamin D3, beta carotene, and dry A vitamin supplement.

On initial examination, Gina showed scoliotic posture with the major curve, a right thoracic curve with a right rib hump (Figure 2). Her sagittal postural deviations included cervical hyperlordosis, thoracic hyperkyphosis, and lumbar hyperlordosis.¹¹ Changes in the frontal plane were a side shift of her thoracic spine to the right and tilting of her neck to the left. Transverse plane changes included right protracted shoulder, left retracted shoulder, and right thoracic rib hump, with neck rotated to right. She had tightness of the hamstring muscles in both legs, a diminished sensation to pain and temperature and impaired gait pattern presenting initial contact on toe in the stance phase in both legs. She also had balance deficits during quick turns and transfers.

Tests and Measures

On direct palpation, Gina had mild to moderate tenderness over her right trapezius and right rhomboids. She did not have any measureable joint limitation in range of motion in her upper and lower extremities. Mild spinal limitation was noted in right side bending and left rotation of her thoracic spine. Gross muscle testing was performed and the patient had a grade of fair for strength in both upper and lower extremities.

Special tests

The Adam's forward bend¹⁰ test was positive for scoliosis with rib hump on the right convexity at thoracic level. Her angle trunk rotation was measured by a scoliometer¹¹ as 6° to 7° at the T8 level. Her Pediatric Balance Test score was 32/56. The Six-minute Walk Test¹² for endurance was 285 m. The 50th percentile for healthy 10-year-old girls is approximately 625 m.¹³ The most recent patient x-ray completed at the time of initial evaluation showed a thoracic Cobb angle^{10,11} of 35°. Table 1 shows further information on all the tests that were performed.

DIAGNOSIS AND PROGNOSIS

Based on information from her subjective history and physical examination, the physical therapist determined that Gina has multiple problems that could benefit from PSSE and conventional physical therapy intervention. Her posture, muscle performance, endurance, and gait pattern limitations should all improve with physical therapy.⁷

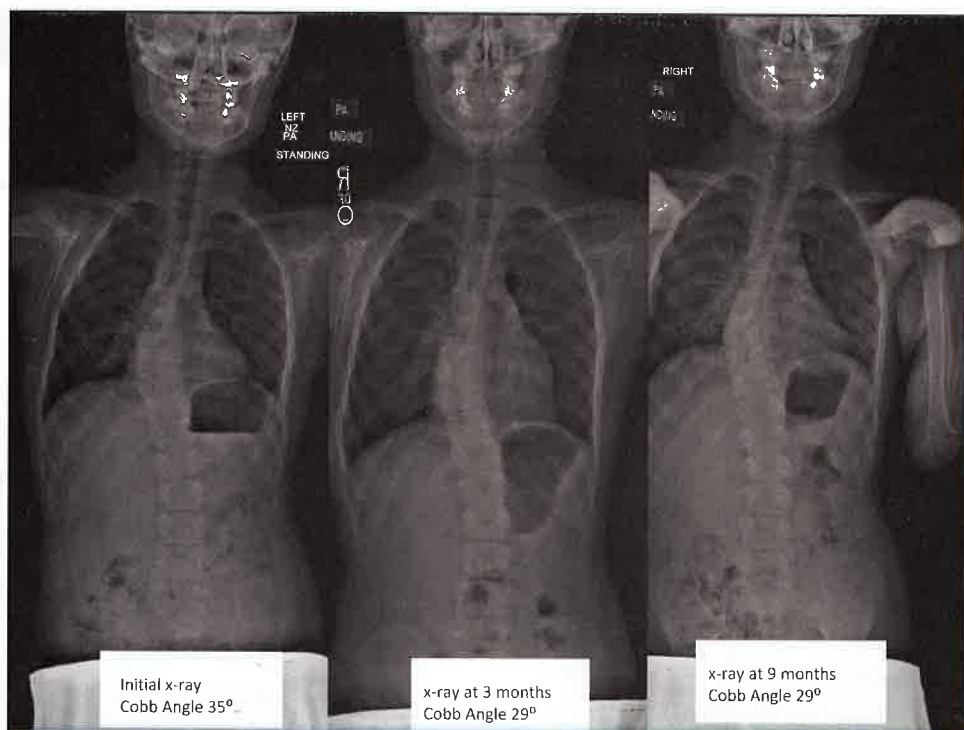


Figure 1. X-rays showing improvement/stabilization of Cobb Angle over the 9-month period.



Figure 2. Image of back. A, initial visit. B, after 9 months.

GOALS

The main goal for outpatient therapy was to control the progression of the scoliotic curve and improve self-image.¹⁰ Gina's parents desired to learn a home exercise program (HEP) and were willing to help their daughter continue with exercises at home. Other

goals were to improve her endurance in community ambulation and increase sports participation. The school-based physical therapy had similar (comparable) goals, with the outcome focused on Gina's ability to participate fully in her educational setting.

Table 1. Treatment Intervention

Months/Treatment	Outpatient	School	Home Exercise Program
0-2	<ul style="list-style-type: none"> - Physical therapy evaluation - Schroth exercises for scoliosis - Active self-correction - Neck stretching and isometric exercises - Manual therapy: trigger point therapy over traps and rhomboids - Gait training (normal heel toe gait) - Family education 	<ul style="list-style-type: none"> - Endurance training for upper extremity and lower extremity - Posture in sitting and standing as per Schroth therapy (positioning at school desk) - Gait training(heel toe) 	<ul style="list-style-type: none"> - Activities of daily living and task oriented, positioning (lying, sitting and conscious posture-self correction as per Schroth therapy) - Neck and hamstring stretches - Schroth exercises - Ergonomic care
2-4	<ul style="list-style-type: none"> - Continued above program - Balance exercises: static and dynamic exercises - Gait training (walking in different directions) 	<ul style="list-style-type: none"> - Strength training exercises with eraBand upper extremity and 2 to 3 lbs for lower extremity - Balance exercises followed in school, one leg standing, open and close eyes activities - Gait training followed 	<ul style="list-style-type: none"> - Continued above exercises - Strength training - Bracing care - Some community ambulation in park to improve endurance and gait pattern
4-6	<ul style="list-style-type: none"> - Continued above program - Advance dynamic balance exercises 	<ul style="list-style-type: none"> - Endurance training-hopping, squatting, step up and down - Balance training on air disc, tandem walking - Gait training-taking turns, upslope 	<ul style="list-style-type: none"> - Continued above exercises - Community ambulation with less fatigue than before
6-8	<ul style="list-style-type: none"> - Schroth exercises - Plank exercises for core strengthening - Progression with strength training 	<ul style="list-style-type: none"> - Core/plank exercises - Endurance exercises-jump rope, crab and bear walking added - Gait training - Strength training using weights - Balance training-dynamic (catching and throwing ball, stepper, obstacle course) 	<ul style="list-style-type: none"> - Continued Schroth exercises - High-endurance activities, ie, jogging, playing with a ball with siblings - Recreational activities (monkey bars, rope climbing)
8-10	<ul style="list-style-type: none"> - Schroth exercises and others continued for maintenance 	<ul style="list-style-type: none"> - High-level strength and endurance program continued 	<ul style="list-style-type: none"> - Continued Schroth therapy - Walked a mile - Participation in playing with siblings - Plays in park on bars and other equipment - Bicycling, ice skating with close supervision/guarding as patient still has some balance deficits

INTERVENTION

Based upon the initial evaluation, a plan of care was developed for Gina that included Schroth therapy, modification of the school physical therapy program to include the Schroth philosophy, and a HEP (see Table 1). Procedural interventions included strength, balance, and gait training to improve general conditioning and endurance. Gina was advised to attend outpatient physical therapy twice a week for an hour, along with a daily HEP. An intervention program was provided with an explanation of technique, and verbal consent was obtained from her parents.

Soft tissue mobilization was performed to improve flexibility, while isometric neck exercises and stretches were performed to improve neck posture. The patient and her parents were instructed in the proper ergonomic care for positioning and lifting in the

home. The patient was given lower extremity strengthening exercises that included squats, lunges (forward and side lunges), and wall push-ups for home and school.¹⁴ Gait training was also performed to improve her heel-to-toe gait pattern. Over the course of treatment, the therapeutic exercise program progressed in intensity and type. Specific exercises were varied to maintain her interest. All exercises were administered under the supervision of her physical therapist and parents. Appropriate levels of verbal and manual cueing were given to maintain her spine in a neutral position. Initially, the patient was given more frequent rest periods. Gina had skin blotching and muscle soreness but over time, with better endurance, these decreased.

The school physical therapy program was modified through a verbal conference between the two therapists. The school thera-

pist agreed to focus on an exercise program to improve strength in the proximal trunk/hip muscles (core) with exercises such as planks, squats, lunges, and modified push-ups. Static and dynamic balance exercises include heel/toe walking, single leg stance on various surfaces, heel/toe raises on a flat surface, and 4" and 6" steps. Additionally, stepping up and stepping down, hopping, jumping rope, turning, throwing, and catching a ball are performed. All exercises were progressive to improve agility and balance.¹⁴

Gina's comprehensive HEP consisted of 30- to 40-minute sessions 5 times a week, divided into 2 components. The exercises consist of Schroth therapy and strength and endurance exercises. The parents are involved in the exercise program and maintain a log of all exercises performed. The family and patient are also educated about postural and

ergonomic care to perform ADLs. Schroth-based ADL exercises are also included to help maintain good posture while performing ADL.¹⁵

Gina showed considerable improvement after 3 months of therapy. Her Cobb angle decreased from 35° to 29° while her core strength increased. Figure 1 shows Gina's x-rays over the 9-month period. Table 2 shows the improvement after 3 months. After 4 months of Schroth therapy, Gina started wearing a night-time Providence brace (Eschen Prosthetics and Orthotics New York City, NY). The Providence brace applies controlled, direct, lateral, and rotational forces on the trunk to move the spine either toward or beyond the midline. The polypropylene plastic brace is fabricated using computer-aided design (CAD) and computer-aided manufacturing (CAM).¹⁶ Her family was educated about the use and risk of bracing and the importance of discipline and adherence to wearing the brace. They were also advised to monitor for any skin redness and lesions.

RESULTS

Gina showed improvement in all areas with therapy. After a few weeks of treatment, there were changes in her overall posture. Her neck posture improved within two months of initiation of Schroth therapy. She does not have any tilting in her neck. She has mild tenderness in her trapezoids and rhomboids, and she still has hyperlordosis in the lumbar area. Figure 2 shows the picture of patient's back at initial visit and after 9 months. Table 2 shows the patient's progress over a 9-month period of outpatient treatment.

One year after the initiation of Schroth therapy, Gina continues to be seen in outpatient treatment one to two times per week. Her mother states that Gina has been more active in outdoor activities, has improved endurance, and does not get fatigued like before. They agree that her quality of life has improved. Her performance and participation are better in both school and recreational activities. Recently she has also started to ride a two-wheeled bicycle. She plays in the park, climbing the monkey bars, and rope climbing nets. She has better balance for ice skating, and she recently participated in a parade in which she walked for a mile. She can run faster with fewer episodes of losing balance while making turns or hopping over an object. She is continuing to progress with her posture and her physical performance.

DISCUSSION

Hayek et al⁹ found that a majority of FD patients develop scoliosis or kyphosis by the age of 10, with a progressive scoliotic curve. Most patients are not able to tolerate bracing due to breathing problems and abdominal difficulties, compliance, and skin pressure ulcers from the brace.⁸ Patients with FD have a low body mass index due to gastrointestinal issues, diminished weight bearing, high bone turnover, and changes in bone profusion, which lead to osteopenia. Surgical procedures, including both posterior spinal fusions with instrumentation and anterior fusions, report a high level of actual and potential problems, including a loss of correction.⁸ These problems in the FD population have resulted in conservative treatment trials rather than early surgical interventions

as seen in idiopathic scoliosis.

As per SOSORT (Society of Scoliosis Orthopedic and Rehabilitation Treatment) guidelines, PSSE are based on the auto-correction of the spine in 3D, training in ADL, stabilizing the corrected posture, and patient education.⁶ One study has shown that active self-correction and task-orientated exercises are superior to traditional exercises in reducing spinal deformities and improving the health-related quality of life in patients with scoliosis.¹⁵ Furthermore, these exercises need to be individualized as per patient's needs, curve pattern, and treatment phase. As recommended by the Scoliosis Research Society, PSSE is commonly prescribed in conjunction with brace treatment or for small curves. Unlike physical therapy, PSSE is less often prescribed for the treatment of pain (only 3%). Instead PSSE is commonly prescribed to improve aesthetics (62%), to prevent curve progression (60%), and to improve the quality of life (53%). Further, the most common specific PSSE used was Schroth (57%), followed by Side Shift (22%), Scientific Exercise Approach to Scoliosis (21%), and Functional Individual Therapy for Scoliosis (19%).⁷ Due to a lack of high-level evidence for the effectiveness of PSSE, a majority of doctors still do not prescribe it.

There is a need for further research on conservative treatment for scoliosis patients as well as FD patients with scoliosis. However, a few studies show the effectiveness of Schroth therapy for scoliosis. A randomized controlled trial study by Kuru et al¹⁷ showed the effectiveness of Schroth treatment in 51 patients with idiopathic scoliosis compared to the control group. The SOSORT guide-

Table 2. Patient's Progress over 9 Months

Test/Time in months	0	3	6	9
Height (in)	48.75	49:25	50	50.5
Weight (lbs)	50	50.5	53	55
X-rays (Cobb Angle degree) Thoracic	35	29	-	29
Angle Trunk Rotation (degree -T8 level) Using Scoliometer	7	5	5	4
MMT Grades (bilateral upper extremity- shoulder, elbow and wrist)	3/5	3+/5	4-/5	4/5
MMT Grade (bilateral lower extremity – hip and knee)	3+/5	4-/5	4/5	4/5
MMT Grade (bilateral lower extremity – ankle)	3/5	3+/5	4-/5	4/5
MMT Grade – abdominals	3-/5	3/5	3+/5	3+/5
Chest Expansion (nipple level)	60 cm	61.5 cm	62 cm	63.5 cm
Pediatric Balance Scale (maximum score-56)	32	35	39	43
Six-minute Walk Test	285 m	305 m	345 m	390 m
Abbreviation: MMT, manual muscle test				

lines offer an actual standard of conservative care, including braces, exercises, sports activities, and assessment. As per the 2011 SOSORT guidelines, 10-year-old patients can start PSSE.¹⁰

Hayek et al⁹ showed limited effectiveness of bracing in patients with FD. As the curve in FD patients is highly progressive, the treatment plan was designed to stop the progression of the curve using a combination of various known conservative treatments. The patient was initially started with Schroth therapy and strength training. A HEP was designed to make sure she was consistent with her exercise routine. Soon, the school therapy program was modified to complement her outpatient treatment. This combination helped to decrease the Cobb angle by 6° in 3 months. She started wearing the night-time brace after 4 months, and still continues the therapy to keep the curve from progressing. The combination of therapy and night-time brace also helps to maintain good posture and may prevent surgery. This case study is unique, as it shows the effectiveness of a combination of conservative treatments and PSSE for scoliosis to improve neuromuscular and orthopaedic problems in FD patients.

CONCLUSION

This case presents a successful intervention for the treatment of scoliosis and other neuromuscular disorders in an FD patient using conventional physical therapy and Schroth therapy for scoliosis. The patient's Cobb angle decreased from 35° to 29° after 3 months and has been stable since then. Schroth therapy and bracing helped to halt the progression of the curve.⁴ Strengthening, balance, and endurance exercises helped to improve the overall physical condition of this FD patient.¹⁰ As the majority of patients with FD will get scoliosis, screening for scoliosis should start early. This will help control the curve in the early stages, using conservative treatment.

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