

Multidisciplinary Approach to Enhancing Function and Quality of Life in Adolescent Rheumatoid Factor Negative Polyarticular Juvenile Idiopathic Arthritis: A Case Study

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Introduction:

Rheumatoid Factor Negative Polyarticular Juvenile Idiopathic Arthritis (RF-negative polyJIA) presents unique challenges in management and rehabilitation, particularly in adolescents like Sara, a 13-year-old girl with an 8-year history of the condition. This case study delves into Sara's medical history, current complaints, physical therapist examination, evaluation and diagnosis, intervention plan, coordination, communication, and documentation, patient education, reexamination, and outcomes. Sara's case highlights the importance of a comprehensive approach to managing polyJIA to improve the patient's quality of life, functional capacity, and treatment adherence.

Case Description:

Sara is a 13-year-old girl with RF-negative polyJIA of 8 years' duration. After several years of mild disease activity, Sara presents with swelling in multiple joints, most notably both knees and the right ankle. She complains of pain, morning stiffness lasting 30 to 60 minutes, and neck pain during the day. She lives with her parents, an older brother, and a sister and is in the eighth grade at a local public school.

Medical History

Sara was diagnosed with polyarticular JIA at 5 years of age. Her parents stated signs of the disease, including stiffness and irritability upon awakening, "bumps" on her elbows and shins, and altered gait pattern, were evident for at least a year before the diagnosis. At her first visit to the pediatric rheumatologist, she had active disease in most extremity joints and the cervical spine. Rheumatoid nodules were found on the extensor surface of the ulna and tibial crest bilaterally. She was originally treated with naproxen; however, after 6 months, during which she continued to have active disease, MTX given orally once a week was initiated. Signs of systemic disease were not evident at this time. During the next several years, Sara continued to have disease flares in both knees, both hips and the right ankle that were managed by IA steroid injections. Following each injection, she reported significant pain relief and was able to return to her normal activities. At the age of 11 years, Sara's JIA was determined to be in clinical remission on medication. However, her disease flared 6 months ago, with increased joint pain, morning stiffness, and fatigue that affected her school attendance and participation in PE and sports.

Current Complaints

Sara is seen in the clinic today to review her medications and HEP. She takes her MTX but misses at least 30% of the prescribed dose of naproxen each week. She admits to poor adherence to the HEP, stating it doesn't help. According to Sara's mother, she has missed school or been late at least once a week for several months owing to disease symptoms. She does not meet the school's PE or sports requirements because of her JIA. She receives Physical therapy and Occupational therapy once a week in school and several accommodations under a 504 plan. These include lockers at either end of the school, a set of books for home, a laptop computer for class notes, extra time for written exams, and an excuse from PE when she is unable to participate. Sara is also not bound by the school's attendance

policy. The rheumatologist confirmed that Sara has active arthritis in both of her wrists, knees and right ankle. Active and PROM is limited in the cervical spine, wrists, hips, right knee, and ankle; she has deformities of the toes in both feet. He added ETN once a week to Sara's medication regimen, continuing the once/ weekly MTX and naproxen twice/day. IA steroid injections were scheduled for her next visit in 1 week, and she was referred to the physical therapist for an evaluation and review of her HEP, and to OT for revision of her wrist splints and suggestions for adaptive equipment to improve her hand function. Sara was also referred to the child life specialist for assistance in managing her own healthcare needs.

Sara's Goals

Sara expressed two major concerns: (1) She would always have this disease and never be able to do the same activities as her friends; (2) She would not be able to keep up physically with the demands of high school. She stated her goals were to be like other kids, play on a sports team like basketball or soccer, and hike and ski with her family.

Physical Therapist Examination

QUESTIONS GUIDING THE EXAMINATION

1. Are there specific activity limitations that negatively impact Sara's participation and overall QOL and prevent her from achieving her current goals?
2. Are there specific impairments that contribute to these problems?

ACTIVITY AND PARTICIPATION FINDINGS

To answer the first question, several standardized assessments validated for children with JIA were used. The QOML questionnaire includes two 100-mm VAS that measure overall QOL and HRQOL; higher scores indicate better QOL. Sara rated both her overall QOL and HRQOL as 40/100 mm. Using the 5-point categorical scale (much better to much worse), Sara rated her life as "much worse" than at her last clinic visit 3 months ago, indicating her JIA hurts her QOL. Sara also completed the CATCHAQ; the Disability Index (DI), calculated as the mean score for all 38 items, has a range of 0 to 3 where higher scores indicate greater disability. Her DI was 1.50, suggesting moderate disability. She scored the following tasks as "unable to do" during the previous week: "play team sports with others in my class," "run a race," and "perform activities for a long time without getting tired." Pain during the previous week was scored as 60/100 mm on a VAS (higher scores indicate greater pain).

To answer the second question, several measures were performed, including a systems review, observational gait analysis, and two composites of the BOT-2, Body Coordination (Balance and Bilateral Coordination) and Strength and Agility (Running Speed and Agility and Strength).

BODY STRUCTURES AND FUNCTION FINDINGS

1. Signs of active joint disease • Both wrists, knees, and ankles had effusions with loss of joint contours; swelling was also noted around the right Achilles tendon. • Tenderness to palpation and mild withdrawal were noted at the above joints and at the right Achilles tendon.
2. ROM: All joints showed PROM within normal limits (WNL) with these exceptions: • Cervical spine rotation and lateral flexion to either side: limited by 50%. • Right shoulder flexion: 220 degrees (0 to 160 degrees) • Elbow extension: 220 degrees (R), 30 degrees (L) • Wrist DF: (R) 225 degrees (0 to 45 degrees); (L) 215 degrees (0 to 55 degrees) • Resting position of R wrist/hand: ulnar deviation, MCP, and PIP flexion (alignment corrected with passive motion) • Pelvis in anterior tilted position; lumbar spine in excessive lordosis • Hip extension (modified Thomas test): 210 degrees (R) and (L). • Ober test: short Tensor Fascia Lata (R) • Knee extension, measured in prone: 210 degrees (R), 25 degrees (L) • Hamstring length test (supine): 245 degrees (R), 235 degrees (L) • Prudential Fitnessgram (PF) Sit & Reach score: 8", below the minimum health fitness standard

(HFS) of 10" for her age and gender
180 • Feet and ankles • (R) ankle PF: 0 degrees to 30 degrees;
(R) ankle DF with the knee extended: 0 degrees • Hindfoot eversion: 0 degrees (R) and (L);
inversion WNL • Pescausus (R) and (L)

3. Muscle strength • Gross strength (MMT): lower limb = 4/5; upper limb = 3+/5 • Functional ankle PF muscle endurance: able to perform eight bilateral heel rises • Grip strength measured with a modified blood pressure cuff: 60/20 mm Hg (R) and 80/20 mm Hg (L). According to Smythe and Helewa,⁷⁴ a rise of 20 mm Hg from the baseline of 20 mm Hg is equal to approximately 5 lb (2.27 kg) of force; Sara's grip strength, measured in pounds, was 10 lb (R) and 15 lb (L), considerably below the reported range of values for healthy, typically developing 13-year old females: (39 to 79 lb [R], 25 to 76 lb [L]).¹⁷⁰ • PF curl-up test (abdominal strength and endurance) score: 8, below the minimum HFS of 18; PF modified pull-up test (upper body strength and endurance) score: 1, below the minimum HFS of 4.¹⁷¹

4. Aerobic performance/exercise tolerance • 6MWD: 550 m, less than mean distance of 663±50.8 m (95% CI: 651.0 to 675.0) reported for 12- to 15-year-old females¹⁷² • Post-walk HR (170 bpm) was approximately 82% of age-related HR_{max} and indicates a good effort and provides a target HR for exercise training • Self-rated fatigue during the previous week: 50 on a 100-mm VAS

5. Body composition: Body mass index and skinfold thickness measure WNL for her age
6. Gait pattern: Footprint analysis with video • Decreased walking velocity: 75 cm/sec compared with 138.8±4.7 cm/sec reported for 12.6 y/o males and females combined • Shortened right step length compared with left • Majority of weight borne on the lateral side of the foot throughout stance phase • Lack of push-off at terminal stance: decreased active ankle PF ROM and decreased hip extension
Evaluation and Diagnosis

GUIDING QUESTIONS

1. Which impairments contribute to Sara's current activity and participation problems?
2. Which impairments must be addressed to prevent or minimize secondary problems?
3. What strengths and resources does Sara have that could support her overall QOL? the impairments believed to contribute to these problems. • Active arthritis contributes to pain, tenderness, and limited active and passive ROM • Limited joint motion and soft tissue shortening contribute to impaired movement patterns, fatigue, and pain during physical activities • Muscle weakness, fatigue, and poor power contribute to gross motor deficits • Impaired joint mobility and muscle performance contribute to gait deviations • Impaired proprioception, coordination, and speed may contribute to poor muscular control and postural stability during challenging physical activities • Poor adherence to prescribed medications and HEP and Sara's limited participation in her health care contribute to poor disease control and functional outcome

STRENGTHS AND RESOURCES

Although Sara's adherence to her medical and therapy regimen has been inadequate, she now appears more interested in managing her disease and improving her health status and functional capacity. Her parents are very supportive, and the school appears to be willing to accommodate her needs by making requested modifications to her educational program. Prognosis

GUIDING QUESTIONS

1. What would improve Sara's active participation in her health care?
2. What is the best POC about medications and rehabilitation?

Question 1: The rheumatology team believed that inadequate disease control and poor adherence to her therapy regimen contributed to Sara's current problems. They thought adherence would improve if Sara were more involved in her health care. Sara stated she wanted to participate in PE and sports with her family and friends. She is also concerned about adjusting to the physical and work

challenges of high school next year. The child life specialist helped her understand how she might achieve her goals if her JIA was under better control by regular use of medication and adherence to her exercise regimen. Question 2: The team believed that Sara's arthritis and functional status would improve following IA injections, the addition of etanercept (Enbrel™), and improved adherence to the NSAID prescription and daily exercise regimen. Sara agreed to a 6-month contract, listing her goals, therapy objectives, and a POC aimed at achieving these. An interim re-evaluation was scheduled for 3 months. The contract included: (1) following her full medication regimen, (2) performing home ROM and strengthening exercises, and (3) participating in an aerobic conditioning program designed by the physical therapist. She also agreed to wear resting hand splints each night while sleeping. Sara and her parents agreed to a 3-month trial of a direct physical therapist and occupational therapist twice a week, with goals of improving ROM and strength, and her parents signed permission for the clinic physical therapist and OT to consult with the school PE instructor to discuss activity modifications so Sara could safely participate in PE. Expected Outcomes (Goals to be achieved in 6 months with progress toward the goals observed at the 3-month follow-up visit):

- Improved adherence to medication should result in improved disease control, reduced joint pain, improved physical function, and HRQOL
- Goal: Sara will show at least a 75% improvement in adherence to her medication schedule based on a daily log, with entries verified by one parent.
- Goal: Sara will demonstrate the improved capability to perform necessary and desired activities based on a reduction of ≥ 0.13 on her self-reported CATCHAQ38 DI.67
- Goal: Sara's self-reported HRQOL score will show improvement based on an increase of ≥ 11 mm in her score on the QOML VAS.
- Sara's gait pattern will improve following joint injections and supportive therapy
- Goal: Sara will demonstrate increased walking speed and symmetry in step and stride length based on observational gait analysis and footprint recording.
- Available evidence suggests that Sara can improve her performance-related physical fitness through a physical conditioning regimen performed twice a week.
- Goal: Sara's 6MWD will increase by at least 48 meters, the minimal detectable change (MDC) reported for healthy children on the 6MWT.97
- Goal: Sara will demonstrate decreased fatigue with physical activity based on self-report using a 100-mm VAS where higher values equal greater fatigue.
- Daily ROM and flexibility exercises should improve joint ROM and soft tissue extensibility, resulting in decreased stiffness and pain.
- Goal: Sara will demonstrate improved passive joint ROM based on goniometric measurement of the shoulders, hips, knees, and ankles.
- Improved physical status should allow Sara to increase her participation in physical activities with her family and friends. Sara will record progress using a daily diary.
- Goal: Sara will actively participate in at least 75% of all PE activities each week, with or without modifications.
- Goal: Sara will participate in at least one recreational PA with family or friends each week for at least 1 hour.

Intervention Plan Following IA injections to the knees, right wrist, and right ankle, Sara was on non-weight bearing for 1 day. After 1 week of modified PA, she resumed all typical activities. Coordination, Communication, and Documentation The rheumatology team worked with Sara and her parents to determine techniques that would improve her adherence to the treatment plan. The nurse provided instruction to Sara and her parents to ensure correct administration of the weekly injections of etanercept and reasons for following the exact prescription for taking the NSAID. The physical therapist provided written and oral instructions, demonstration, and illustrations of Sara's exercises. The OT made new resting wrist/hand splints and gave Sara assistive devices to improve her ability to perform ADLs. The child life specialist helped Sara establish a schedule for taking medications and develop a daily diary to keep a record of her medications, use of splints, HEP, PE, and recreational activities. Each section of the diary included space for Sara's comments. She was encouraged to contact the team with questions about her program or symptoms. The team, with the permission of Sara and her parents, sent a copy of the physical therapist and occupational therapist evaluations to the school and requested the PE instructor to consult with the clinic physical therapist to adjust activities so that Sara could increase her participation. Patient Education The clinic physical therapist discussed the findings of the

physical examination with Sara and her parents. She discussed the impact of arthritis on joints and muscles, the potential sources of pain and stiffness, and secondary problems, including poor exercise tolerance and impaired motor skills. She explained that Sara's current functional problems were likely related to limited joint mobility and soft tissue extensibility, pain, and poor aerobic and muscular fitness. She explained that intervention would be directed toward reducing these impairments. The therapy program would also include practice of difficult activities for Sara.

Procedural Interventions • Direct physical therapy 30 to 45 minutes twice a week and instruction in HEP.

INTERVENTIONS FOR IMPAIRED JOINT MOBILITY AND MUSCLE FUNCTION

Direct therapy: Initial instruction in maintaining control of the trunk and lumbar spine by engaging abdominal muscles during all activities

1. AAROM for shoulder flexion, abduction in the scapular plane, and medial and lateral rotation, with attention to scapular position, movement, and scapulohumeral rhythm
2. Prone scapular progression exercises; progress to standing scapular exercises against a wall; progress to using light handheld weights when Sara can perform exercises without pain or compensatory movements
3. AAROM and AROM for serratus anterior; progress to exercise against resistance
4. Instruction in isometric exercise to improve neck stability
5. Stretching of shortened latissimusdorsi, lateral shoulder rotators and posterior capsule, hip flexors, hamstrings, and right gastrocnemius using autogenic and reciprocal inhibition
6. Stretching of right TFL
7. Strength training for hip extensors, deep external rotators, and abductors • Begin with AROM to teach correct technique for each muscle group • Progress to graded resistance exercise using light weights or elastic bands • Closed kinetic chain (CKC) exercise, including graded squats, lunges, and "step" training to improve strength, endurance, and control of lower extremity musculature
8. Gait training to improve lower limb weight bearing, step and stride length

INTERVENTIONS TO ADDRESS IMPAIRED MOTOR SKILLS

1. Activities to improve proprioception, agility, and coordination once joint ROM and muscle strength improve and pain decreases
2. Gait training to increase walking velocity, using timed walks
3. Activity- or sport-specific training to ensure safe participation in physical activities and recreational sports

HOME EXERCISE PROGRAM

1. Daily AROM exercises concentrating on cervical spine, shoulders, wrists, hips, knees, and ankles using illustrations from *Raising a Child with Arthritis*
2. Daily aerobic activity (Sara's choice), gradually increasing the duration to at least 30 minutes a day and intensity to at least 75% of her age-based HRmax

RECOMMENDATIONS/REFERRALS

1. Referral to orthotist for custom insoles to accommodate pescausus deformity and decrease pain under MTP joints
 2. Recommendation for semi-rigid cervical collar when riding in school bus and car, and easel top for desk to decrease neck strain when reading
- Re-examination Sara was seen by the clinic physical therapist at her 3-month follow-up rheumatology appointment. She reported receiving physical therapy twice a week at school before classes begin and OT once a week during school time. She also attends all PE classes weekly and participates in approximately 50% of the activities with some modifications. The school PE instructor and physical therapist are working together to improve Sara's gross motor skills and adapt difficult activities to allow her to participate with her classmates.

FINDINGS OF REEXAMINATION

Activities and Participation

1. Sara's CATCHAQ38 DI decreased from 1.50 to 1.30, exceeding the reported minimal clinically important improvement of ≥ 0.13 . Her self-reported HRQOL improved from 40 to 70 mm, and she reported her life as somewhat better since her last clinic visit Body Structures and Functions. Signs of active joint disease • Mild swelling noted in the right knee, no tenderness or pain on passive motion. ROM, flexibility, and joint alignment • No change in cervical spine AROM, but no c/o POM • (R) shoulder PROM: 0 to 170 degrees with stress pain at end range • Wrist DF: 0 to 60 degrees (R), 0 to 60 degrees (L) • Passive hip extension limitation: 5 degrees • Knee extension (examined prone): (R); WNL (L) • (R) Ankle PROM: PF = 0 to 40 degrees; DF with knee extended = 0 to 5 degrees • Passive calcaneal eversion increased: 0 to 5 degrees 3. Standing posture • Sara continued to stand with an increased anterior pelvic tilt and lumbar lordosis, although she could correct this upon request by engaging her abdominal muscles. • Barefoot, Sara continued to stand with most of her weight over the lateral border of her foot; when wearing her custom-made in-shoe orthoses and sneakers, her weight was borne more evenly over the plantar surface of the foot. 4. Muscle strength and flexibility • Grip strength: 100/20 mm Hg (20 lb) (R); 140/20 mm Hg (30 lb) (L) • Score on the PF reported by Sara's school physical therapist improved • Curl-up test: increased from 8 to 12 (HFS = 18) • Modified pull-up test: increased from 1 to 5 (HFS = ≥ 4) • Sit & reach test: increased from 8" to 9" (HFS = 10") 5. 6MWD increased from 550 m to 610 m, exceeding the MDC of 48 m. 6. Gait velocity increased from 75 cm/sec to 110 cm/sec. Gait pattern showed improved heel contact, weight more evenly distributed over the plantar surface of the foot during mid-stance, improved push-off on the medial side of the forefoot and hip extension at terminal stance, and increased step length.

Outcomes

1. A review of Sara's medication and activity diary indicated improved adherence to her medication regimen and HEP program; this was supported by Sara's parents as well as decreased signs of active arthritis and improved ROM in most joints.
2. Improvements in self-reported worst pain from 60 mm to 40 mm and worst fatigue from 50 mm to 20 mm over the previous week on the VASs reflect improved disease control.
3. Improvements in PF scores, walking speed, and 6MWD suggest increased stamina, as a result of improved adherence to her exercise program.
4. Clinically meaningful improvements in her physical function and QOL lend support to her improved health status and ability to manage her condition. Plan Based on Sara's improved physical status, the clinic therapist recommended continuing with direct physical therapy in school with an emphasis on increasing aerobic fitness, strength, and gross motor proficiency. She also suggested that the school physical therapist and the PE instructor work together with Sara on specific training for basketball to allow her to participate in an intramural league at school. A re-evaluation was scheduled for her next clinic appointment in 3 months.

References:

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