

POSITION STATEMENT

PHYSICAL ACTIVITY RECOMMENDATIONS FOR CHILDREN WITH SPECIFIC CHRONIC HEALTH CONDITIONS: JUVENILE IDIOPATHIC ARTHRITIS, HEMOPHILIA, ASTHMA, AND CYSTIC FIBROSIS

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INTRODUCTION

Sports medicine pioneers, such as Oded Bar-Or, recognized the need to study exercise risks and benefits in children with chronic disease.¹ The Canadian Paediatric Society (CPS) and Canadian Academy of Sport Medicine (CASM) endorse the present statement regarding issues in children with juvenile idiopathic arthritis, hemophilia, asthma, and cystic fibrosis.

JUVENILE IDIOPATHIC ARTHRITIS

Juvenile idiopathic arthritis (JIA) (previously known as juvenile rheumatoid arthritis) has a prevalence of 1 in 1000² children. There are 7 subtypes of JIA, likely representing different pathogenic mechanisms.³ Chronic arthritis is defined by the presence of joint swelling or by 2 or more of the following: joint pain, warmth, and limited range of motion for at least 6 weeks. Constitutional signs or symptoms include anorexia, weight loss, growth failure, and fatigue. Extra-articular manifestations include ocular, cardiac, pulmonary, and hematopoietic involvement. Juvenile idiopathic arthritis persists into adulthood in up to 55% of patients, which may have a major impact on physical or psychosocial function. Children with JIA have reduced vigorous physical activity levels, sports participation, and decreased fitness.⁴

Muscle atrophy, weakness, and anemia contribute to reduced fitness, but deconditioning from reduced physical activity is likely the greatest cause. Reduced participation because of disease symptom severity, treatment-related side effects, or worries that exercise may aggravate disease are problematic.

Potential Benefits of Exercise

Physical activity should be encouraged in all children. The psychosocial benefits of group participation cannot be understated. Exercise can have positive biological effects by reducing loss of proteoglycans and cartilage damage, and optimizing bone mineral density. Active children have lower obesity risks, which can worsen joint load.

Research suggests children with JIA can participate in aquatic or land-based weight-bearing exercise programs without disease exacerbation.⁴ Aquatic exercise encourages range of motion, strength and fitness, with less stress on joints. Weight-bearing activity helps promote bone health.

Most published studies are small, not randomized, have great variation in study design and use different exercise modalities. The exercise intensity, frequency, and duration also vary. Despite these differences, the results are generally consistent, suggesting an exercise program (a minimum 6 weeks) improves aerobic fitness; allows better muscle strength and function; decreases disease activity; improves self-efficacy, energy level, and quality of life; and reduces pain and medication use, with no clear effect on function during activities.⁴⁻⁸ Importantly, sports participation does not appear to exacerbate disease.⁹ The 2002 Exercise and Physical Activity Conference Arthritis Working Group guidelines recommend moderate fitness and strengthening exercises for children with JIA.⁸

Potential Risks of Exercise

Conventionally, affected children were advised to limit strain on arthritic joints for fear it may aggravate joint pain or swelling, increasing risk of injury. Muscle atrophy surrounding active joints and periarticular osteopenia may increase the risk of fracture. The effect of tissue loading during exercise on joint surfaces and growth plates in children with arthritis is unknown, and requires further study.



Young children with JIA may have gross motor delays, affecting sport readiness. Children with cervical spine arthritis are at greater risk of spinal cord injury (especially during contact sports), and those with temporomandibular joint disease may sustain dental injury. Complications of JIA, such as uveitis and its sequelae (visual impairment), may increase the risk of eye injury. Myocarditis and pericarditis in systemic arthritis, and aortic valve insufficiency or aortic root anomalies in HLA-B27-associated arthritis, may increase the risk of cardiovascular complications with exercise.¹⁰

Children with long-standing JIA may have difficulties with endurance sports. Greater submaximal energy expenditures are reported, suggesting increased metabolic demands for routine physical activity.¹¹ A meta-analysis of 5 JIA studies found that aerobic fitness in children with JIA was 22% lower than their healthy peers.¹² Most research suggests aerobic fitness is not related to disease severity or activity but rather to disease duration.¹²⁻¹⁴

Recommendations: Children with Juvenile Idiopathic Arthritis (JIA)

- * Can safely participate in sports without disease exacerbation.
- * Should participate in moderate fitness, flexibility, and strengthening exercises.
- * Can participate in impact activities and competitive contact sports if their disease is well controlled and they have adequate physical capacity.
- * Should be encouraged to be physically active as tolerated. Those with moderate to severe impairment or actively inflamed joints should limit activities within pain limits.
- * Should gradually return to full activity following a disease flare.
- * Should take individualized training (especially for children with severe joint disease) within a group exercise format for physical/social benefit. Physiotherapists on pediatric rheumatology health care teams should coordinate individual exercise programs.
- * Should have radiographic screening for C1-C2 instability before participation in collision/contact sports if they have neck arthritis. If present, further evaluation is required.

* Should wear appropriately fitted mouth guards during activities with jaw and dental injury risk (per general population), especially if they have jaw involvement.

* Should wear appropriate eye protection (per general population) during activities with ocular injury risk.

HEMOPHILIA

Hemophilia is an X-linked recessive inherited bleeding disorder caused by the absence, deficiency, or dysfunction of plasma coagulation factor VIII or IX. Hemophilia has an incidence of 1 per 5000 newborns. Clinical phenotype and risk of hemorrhage varies from mild to severe, and is related to functional plasma factor levels.¹⁵

Children with severe hemophilia (< 1%-2% of normal factor levels) have spontaneous bleeding even without trauma. Joint or muscle hemorrhage, easy bruising, and prolonged bleeding after trauma are common, and severe bleeding (intracranial, vital organs, airway) may occur. Repeated joint hemorrhage causes synovitis, leading to joint degeneration and arthropathy. Hemophilia is characterized by joint contractures, limited range of motion, and chronic pain. The knees, ankles, and elbows are most commonly affected. Prophylactic treatment with the deficient factor reduces spontaneous bleeding and the risk for hemophilic arthropathy.

Potential Benefits of Exercise

Regularly active children at hemophilia camps have fewer bleeding episodes than their sedentary peers.¹⁶ Greater muscle strength around affected joints may help to protect joints from hemarthrosis, increase joint stability, and reduce injury risk.¹⁷ Prophylactic physical therapy improving periarticular muscle strength was demonstrated to reduce the frequency of hemorrhage.^{17,18} Proprioceptive training may decrease joint damage and improve athletic performance.¹⁹ Weight-bearing exercise can improve bone health in children with severe hemophilia who have reduced bone mineral density.²⁰ Aerobic exercise may have a beneficial effect on coagulation. Vigorous exercise increases factor VIII levels transiently in healthy individuals, and submaximal exercise can modify coagulation parameters in those with mild to moderate hemophilia.²¹

Potential Risks of Exercise

Fitness, anaerobic power, and muscle strength is lower in children with hemophilia.^{19,21} Affected children may restrict activity due to parental concern, musculoskeletal pain, or deconditioning. Although hemophilia itself does not negatively affect fitness or athletic performance, chronic hemophilic arthropathy may lead to impaired neuromuscular function, diminished muscle strength, and endurance. Participation in collision or contact sports can result in a life-threatening bleeding event. The actual risk of muscular, articular, and intracranial hemorrhage depends on the individual child's hemorrhagic tendency, history of bleeds, prophylactic treatment, and sport participation.

Recommendations: Children with Hemophilia

- * Should receive appropriate factor prophylaxis to reduce the risk of bleeding in sport.
- * Should undergo vigilant assessment of joint and muscle function before sport selection. If restrictions are required, physicians should counsel children and their families about safe alternatives.
- * Should be carefully assessed before allowing participation in contact or collision sports such as martial arts, hockey, or football. Consultation with a sport medicine physician and/or pediatric hematologist may help.
- * Require written strategies (coach, parent, or school) before sport participation to prevent or treat bleeds.
- * Should wear protective equipment, undergo physical therapy, or take prophylactic factor replacement therapy.
- * Require factor replacement, ice, splinting, and rest to manage acute bleeds. Physical activity should be avoided until joint pain or swelling has resolved. Return to sport requires individualized assessment and appropriate rehabilitation.

ASTHMA

Asthma is the most common pediatric chronic disease, with over 300 000 Canadian children affected.²² Higher rates appear to be associated with poor socioeconomic status, obesity, and low physical activity levels. Asthma is a chronic inflammatory disorder of the airways, characterized by airway hyperresponsiveness and reversible airflow limitation. Typical presenting symptoms include shortness of breath, cough, and wheezing. Asthma is associated with greater bronchial hyper-reactivity to viral infections, cigarette smoke, inhaled allergens, emotional stress, environmental factors, and exercise. Exercise and emotions trigger bronchospasm but minimal inflammation. Rarely is exercising the only trigger, making 'exercise induced bronchospasm' (EIB) better terminology.

Almost 90% of asthmatics and 40% of individuals with allergic rhinitis experience EIB. In children, EIB may be the first presentation of asthma. Overall prevalence in high school, college, and Olympic athletes is 12%,²³ although this is probably underestimated.²⁴ Exercise-related dyspnea is often mistakenly diagnosed as EIB; however, bronchial hyperresponsiveness is not associated with exercise-related dyspnea.²⁵

In patients with EIB, bronchoconstriction typically occurs after 8 to 15 minutes of physical activity and resolves within 60 minutes. Running and other land-based cardiovascular exercise (rarely swimming) are common triggers. One popular hypothesis regarding EIB pathogenesis is the evaporation of water lining the airways secondary to higher ventilatory rates during and after exercise.²⁶ Alternatively, cold, dry air may cause an osmotic gradient across mast cells, resulting in mediator release. Fifty percent of asthmatic children without EIB symptoms can be diagnosed using an exercise challenge pulmonary function test (PFT).^{26,27} A drop of 10% to 15% in forced expiratory volume (FEV₁) in 1 second from baseline, following vigorous exercise for approximately 6 to 8 minutes, is diagnostic of EIB. The sensitivity and specificity of exercise PFT in children are up to 63% and 94%, respectively.²⁸ Eucapnic voluntary hyperventilation testing is the preferred challenge test for EIB in athletes, because pharmacological challenge tests have low sensitivity in this setting.²⁹

Asthma management should include identification of disease severity and known triggers, and the creation of a written action plan. Those with persistent symptoms and/or abnormal baseline PFT require ongoing anti-inflammatory treatment with inhaled corticosteroids and/or leukotriene antagonists. Beta₂-agonists are used as rescue medication or before exercise to prevent EIB.

Children with mild intermittent disease triggered by exercise may benefit from non-pharmacologic interventions (nose breathing and warm-up exercises) as well as pre-event inhaled beta₂-agonists. Those in prolonged activities may benefit from a long-acting beta₂-agonist (formoterol) with rapid onset. Athletes should take these agents 15 to 30 minutes before exercise.³⁰ Those who compete nationally and internationally require a therapeutic use exemption form, with documentation of asthma or EIB to use certain medications.^{27,30} Athletes are strongly advised to consult the Canadian Centre for Ethics in Sport (www.cces.ca), the World Anti-Doping Agency (www.wada-ama.org), and their international sport federation to determine the current required documentation.

Children with asthma exhibit similar activity levels as unaffected peers.³¹ Both have similar self-perceptions or physical self-concept. Disease severity and parental concerns present possible barriers.

Potential Benefits of Exercise

Physical and/or psychosocial benefits of exercise are evident. Bronchial hyperresponsiveness increases with decreasing hours of exercise per week.³² Swimming can increase aerobic fitness and decrease asthma morbidity.³³ Exercise training can improve aerobic capacity; however, PFTs do not change significantly.³⁴

Exercise may decrease EIB severity by increasing the threshold for triggering bronchospasm. Almost 50% of affected individuals can experience this 'refractory period' up to 4 hours after initial exercise, resulting in decreased bronchoconstriction during subsequent exercise.³⁵ In some cases, athletes can warm-up with exercise 45 to 60 minutes before scheduled activities to reduce their subsequent asthmatic symptoms, and to improve exercise capacity and quality of life.

Potential Risks of Exercise

High-intensity exercise can trigger EIB by increasing minute ventilation and respiratory heat/water losses leading to a greater drop in forced expiratory volume FEV₁.³⁶ Permanent bronchial changes may occur in endurance athletes, who seem to have higher rates of bronchial hyperresponsiveness.³⁷ Certain sports expose individuals to dry, cool air,³⁸ environmental allergens, and pollutants, which may trigger flares. Athletes in running and winter sports have more reported symptoms.³⁹ Breathing humid air during swimming may be protective,⁴⁰ but

potential risks from exaggerated parasympathetic tone ('diving reflex'), and chlorine-related airway irritation that triggers bronchoconstriction may occur.³³ It is controversial whether asthma patients are at higher risk of scuba diving injury. They should have normal spirometry (especially residual volume) at rest and in response to exercise before being certified to dive.

Asthma-related deaths of individuals younger than 20 years of age, although rare, have been reported in both competitive and recreational sports (eg, basketball and track).⁴¹

Recommendations: Children with Asthma

- * Are able to participate in any physical activity if symptoms are well controlled. Swimming is less likely to trigger EIB than running.
- * Should keep an accurate history of symptoms, trigger exposures, treatments, and course of recovery from episodes of bronchospasm.
- * Should be diagnosed with EIB by a drop in FEV₁ (10%-15%) after a 6 to 8 minute exercise challenge and a positive response to beta₂-agonist medication. Eucapnic voluntary hyperventilation testing is recommended in athletes.
- * Should use leukotriene inhibitors, inhaled corticosteroids, and/or long acting beta₂-agonists for optimal long-term disease control, and avoid overuse of short-acting beta₂-agonists.
- * Should take inhaled beta₂-agonists 15 to 30 minutes before exercise.
- * Should not scuba dive if they have asthma symptoms or abnormal PFTs.
- * Who compete nationally or internationally require a therapeutic use exemption with confirmation of asthma and/or EIB to use certain medications. Consultation with a sport medicine physician is suggested.

CYSTIC FIBROSIS

Cystic fibrosis (CF) is the most common lethal autosomal recessive disease in Canada, affecting 1 in 3600 Caucasian live births.⁴² Men and women are equally affected, although men have a longer life expectancy.⁴³ Cystic Fibrosis is caused by mutations in the CF transmembrane conductance regulator protein, a complex chloride channel located in all exocrine tissues.⁴⁴ Abnormal chloride transport leads to viscous secretions in the pulmonary, gastrointestinal, endocrine, and reproductive systems, and greater salt loss in sweat. Diagnostic sweat chloride testing remains the gold standard. Sixty percent are diagnosed by 1 year of age and 90% by 10 years of age.⁴² Rapid diagnosis using genetic and newborn sweat chloride show promise.⁴² Pulmonary disease causes most morbidity and mortality, but early diagnosis and improvements in therapy have increased mean survival rates to 33 years of age.⁴⁵

Physicians emphasizing exercise, in addition to routine CF treatment, help CF children develop positive attitudes toward exercise. Some may become triathletes or marathon runners.⁴⁶ Disease severity is variable among children with CF, affecting individual exercise tolerance.

Potential Benefits of Exercise

Cystic fibrosis children with high aerobic fitness experience slower deterioration in lung function and greater survival rates.⁴⁷⁻⁴⁹ Training programs can improve exercise tolerance, particularly those with low fitness levels.⁵⁰ Enhanced lung mucous clearance can occur during intense exercise.⁵¹ Swimming, walking, and jogging can improve strength and endurance of respiratory muscles.⁵² Strength training may improve fat-free mass, weight gain, muscle strength, and FEV₁ in affected patients.⁵³

Potential Risks of Exercise

Children with CF may cough with exercise, causing brief oxygen desaturation. However, there is no evidence this effect causes significant injury.^{46,54} Some children cough because of underlying asthma. Major limitations to exercise are degrees of lung disease and subnormal ventilatory capacity. These limitations may be a consequence of bronchial narrowing (edema), bronchospasm, mucous plugging, and reduced alveolar ventilation.⁵⁵ Lung parenchyma destruction results in decreased diffusing capacity leading to oxygen desaturation, CO₂ retention, and cyanosis.^{55,56} Desaturation of arterial oxygen from significant ventilation-perfusion

mismatching, intrapulmonary right-to-left shunting or cor pulmonale with congestive right heart failure⁵⁷ occurs. Cardiac dysfunction is noted in patients with advanced CF (resting FEV₁ lower than 50% predicted) who have lower stroke volumes or cardiac output, and in mild CF patients during submaximal exercise testing. Maximal heart rate during testing is often lower than healthy peers.⁵⁸ All CF patients can develop localized air trapping, increasing the risk of air embolus or pneumothorax during scuba diving.

Resting energy expenditure is 5% to 25% higher in CF youth,⁵⁹⁻⁶² limiting exercise tolerance. Chronic malnutrition may cause lower muscle mass or strength (respiratory or skeletal), impairing sport performance.⁶³ These working muscles have poor oxidative efficiency, contributing to early fatigability.⁶³

Affected children have greater sweat-related salt losses, making exercise in hot or humid environments challenging.⁶⁴ Prolonged exercise (1.5-3 hours), can lead to hyponatremic dehydration.⁶⁵ Prevention by ingesting flavored sodium chloride-containing drinks (50 mmol/L) above thirst levels is recommended.⁶⁵ Cystic fibrosis-related diabetes mellitus make hypoglycemia and dehydration (polyuria) potential concerns with prolonged exercise; hence, additional carbohydrate supplementation is required.⁶⁶ Multilobular biliary cirrhosis and portal hypertension are frequent complications of CF liver disease, leading to esophageal varices and splenomegaly. Those with splenomegaly or liver dysfunction have a higher risk of organ damage during contact or collision sports.

Recommendations: Children with CF

- * Should be encouraged to participate in any physical activity. Consultation with a sport medicine physician or pediatric respirologist is suggested.
- * Should have individualized exercise programs that include strength training.
- * Require supervised or unsupervised home exercises that elevate heart rate 70% to 80% of maximum to increase aerobic exercise tolerance.
- * Who cough during exercise should not necessarily stop activity.



* Those with severe CF should undergo exercise testing to identify maximal heart rate, levels at which oxygen desaturation and ventilation limits occur, exercise-related bronchospasm, and response to therapy.

* Should absolutely avoid scuba diving.

* Should drink flavored sodium chloride-containing fluids above thirst to prevent hyponatremic dehydration. Those with diabetes mellitus require additional carbohydrates during prolonged exercise.

* With an enlarged spleen or diseased liver should avoid contact or collision sports.

CONCLUSION

Physical activity and sport are primary means of exercise and social activity in childhood. Through participation, children can develop fitness, social skills, and relationships. Despite chronic disease, each individual has a unique exercise tolerance and physical capacity.

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The recommendations in this statement do not indicate an exclusive course of treatment or procedure to be followed. Variations, taking into account individual circumstances, may be appropriate. All Canadian Academy of Sport and Exercise Medicine position statements are reviewed, revised, or retired as needed on a regular basis.

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