

SPORTS CARDIOLOGY AND THE PARALYMPIC ATHLETE

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The care of elite athletes with congenital and acquired physical disabilities involves much more than understanding the physical adaptations, altered biomechanics and patterns of musculoskeletal conditions that result from the specific stresses of their sport. Medical professionals who care for Paralympic athletes or provide medical coverage during Paralympic events must be familiar with the changes to the cardiovascular (CV) system that can occur as a result of disability. Conditions unique to certain disability patterns include autonomic dysreflexia, orthostatic hypotension, hypertension and structural changes to the myocardium. These CV risks must be considered when co-ordinating the periodic health evaluation and event coverage for Paralympic sports.

PREVALENCE OF CV ABNORMALITIES IN PARALYMPIC ATHLETES

Athletes with congenital disabilities appear to have a higher prevalence of underlying CV disorders when compared

to athletes in the general population. The combined prevalence of all CV disorders known to cause sudden cardiac death (SCD) in the young athletic population is estimated to be 3 cases per 1000 persons. Hypertrophic cardiomyopathy and autopsy-negative sudden unexplained death are the lead findings associated with SCD¹. Comparatively, in a recent study of 267 Paralympic athletes, structural CV abnormalities were identified in 33 athletes (12%), with 2% demonstrating high-risk for SCD. Among these athletes, cardiac symptoms were not endorsed and physical exam findings were unremarkable. Additionally, of the 105 athletes with 6-year follow-up data, 6% developed CV diseases. Although the increased age of the population (mean age of 36 in men and 34 in women) may have played a role in the higher prevalence of disease, studies such as these suggest a need for pre-participation screening with specifically tailored recommendations for certain impairment groups of Paralympic athletes².

EFFECTS OF PHYSICAL DISABILITIES ON THE CV SYSTEM

Spinal cord injuries – chronic effects

Athletes with cervical spinal cord injuries (SCI) have been found to have attenuated left ventricular mass, dimensions of the left atrium and ventricle, and inferior vena cava³. These structural changes are hypothesised to be the result of relative decreases in cardiac output, stroke volume, cardiac preload, systolic blood pressure (SBP) and heart rate (HR). In a cross-sectional study comparing 44 elite hand cyclists with SCI to 19 non-SCI blind/visually impaired tandem cyclists, no difference in left ventricular function was seen despite SCI athletes having lower LV mass, posterior wall thickness, interventricular wall thickness and left atrial volume⁴.

Additionally, individuals with SCI are at increased risk of coronary heart disease (CHD) and metabolic syndrome compared to the general population. These co-morbidities are attributed to lower energy expenditure from motor function limitations and

fewer opportunities for exercise⁵. However, consistent high-level physical activity in those with SCI can maintain insulin sensitivity to the same level as sedentary able-bodied (AB) individuals and perhaps even improve insulin sensitivity and dyslipidaemia compared to the AB controls⁶.

Spinal cord injuries – autonomic dysreflexia

Athletes with SCI at or above the T6 spinal cord level are at risk of developing autonomic dysreflexia (AD), a potentially life-threatening condition resulting from excessive sympathetic nervous output due to uncontrolled spinal reflexes. Without prompt recognition and intervention, AD causes extreme hypertension that can exceed 200/100 mmHg and may lead to myocardial infarction, intracranial or retinal haemorrhage, seizure, pulmonary oedema, renal insufficiency and even death⁷⁻⁹.

If there is a strong noxious stimulus below the T6 spinal cord level, peripheral nerves attempt to send a signal via the spinal cord to the brain. However, this signal is blocked at the level of the spinal cord injury (Figure 1). Simultaneously, a massive reflex sympathetic surge from T1 to L2 is activated, resulting in vasoconstriction of the blood vessels and a rapid rise in systemic blood pressure (BP). Carotid and aortic baroreceptors detect this uninhibited rise in BP and send a signal to the brain. As the brain tries to send inhibitory impulses to the sympathetic outflow levels in the thoracolumbar spine, these impulses are blocked at the level of the SCI. The remaining compensatory response by the brain is to signal the heart to slow the HR to try to decrease the BP via cranial nerve X, the vagus nerve. Additionally, the blood vessels above the level of SCI can vasodilate. However, these measures are inadequate and the systemic hypertension continues.

The severity of AD is directly related to the SCI level and severity of SCI, with higher SCI levels and neurologic complete injuries at greatest risk. Anatomically, the T6 spinal cord level is important because the greater splanchnic nerve (T5-9) controls the splanchnic vascular bed, which is one of the body's largest reserves of circulatory volume. SCI at or above the T6 level allows the uninhibited sympathetic reflex to constrict the splanchnic vascular

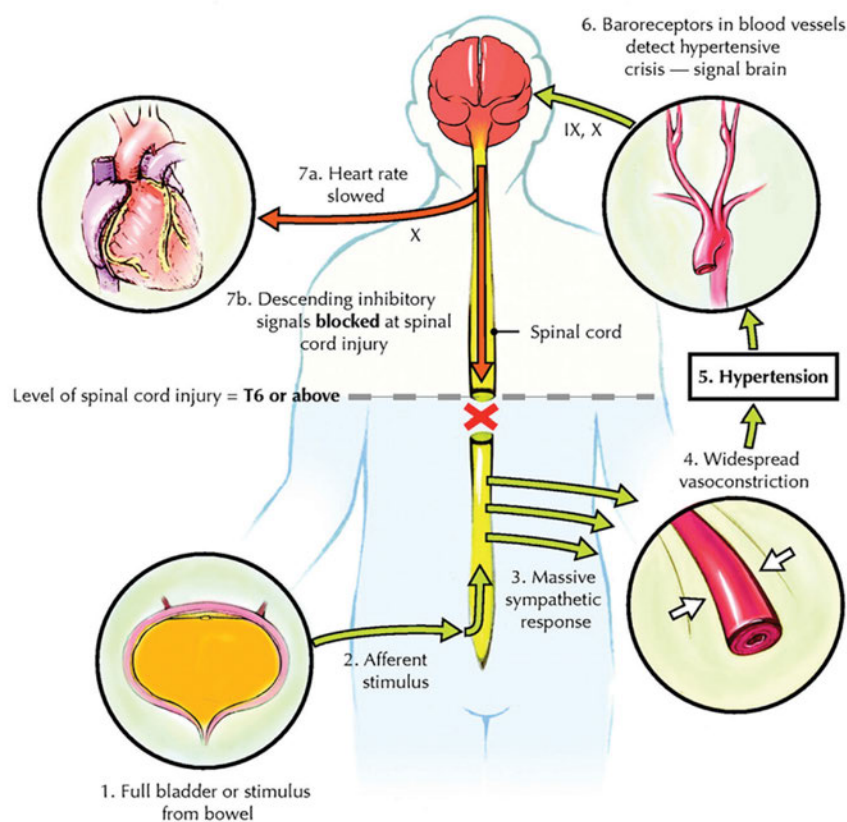


Figure 1: Pathophysiology of AD. Reproduced with permission from Blackmer (2003)²⁹.

bed, causing systemic hypertension. Comparatively, athletes with SCI below T6 usually allow enough descending inhibitory parasympathetic control to modulate the tone of the splanchnic vascular bed and thus prevent hypertension.

Spinal cord injuries – orthostatic hypotension

Impaired autonomic control affects athletes with SCI beyond the risk of AD. Individuals with cervical and high-thoracic SCI have comparatively lower resting BP at baseline and attenuated responses to elevate HR and auto-regulate BP during training and competition compared to those without SCI. Haemodynamically, this results in decreased venous return, stroke volume and cardiac output^{9, 10}. Orthostatic hypotension is also an issue for athletes with cervical and high-level thoracic SCI due to the loss of sympathetic-mediated vasomotor tone, leading to blood pooling in the periphery and splanchnic vasculature. As a result, symptoms of orthostatic hypotension include fatigue, light-headedness, nausea, blurred vision and syncope.

Amputees – chronic effects

In athletes with congenital limb deficiencies or acquired limb amputations,

there can be alterations in vascular physiology proximal to the site of amputation. Specifically, changes in vascular shear stress and circumferential strain in amputees have been hypothesised to contribute to the development of atherosclerosis and arterial remodelling as well as hypertension, LVH and CHD11. Coronary artery calcification scores have also shown to be elevated in amputees, with even higher scores in traumatic compared to dysvascular amputees¹². Post-traumatic lower limb amputees have increased morbidity and mortality from CHD, with proximal leg amputees at greater risk than distal and bilateral at greater risk than unilateral. Similar to athletes with SCI, the increased risk of CHD and metabolic syndrome in amputees may be reduced by high levels of physical activity.

Cerebral palsy

Individuals with cerebral palsy (CP) have been found to have decreased heart rate variability, suggesting a sympathovagal imbalance. Compared to sibling volunteers, ECGs of those with CP have an increased heart rate and mean corrected QT interval, which could result in an increased risk of cardiac arrhythmias and death. Additionally,

snoring is more prevalent among individuals with CP, likely due to upper airway and facial muscle tone alterations and may be indicative of obstructive sleep apnoea which is associated with acute and chronic CV changes¹³.

Stroke (cerebrovascular accident)

Cardiovascular disease has been reported in up to 75% of stroke survivors. As exercise places greater cardiac demands on hemiplegic athletes compared to able-bodied athletes, those with pre-existing cardiovascular disease may have an increased risk for exertion-related cardiac complications¹⁴. Few studies have defined the cardiorespiratory response to exercise in the general stroke population. Specifically, when compared to able-bodied subjects, those with stroke show lower heart rate and blood pressure responses to progressive physical activity¹⁵.

Short stature (achondroplasia)

This autosomal-dominant skeletal dysplasia is associated with narrowing of the foramen magnum and jugular foramina due to diminished growth at the base of the skull. The associated venous narrowing can initially result in increased intracranial venous pressure and later hydrocephalus and ventriculomegaly. The foramen is also anteriorly placed, causing hyperextension

of the brainstem and cervicomedullary compression. These factors, along with mid-facial hypoplasia and small thorax, can lead to cardiorespiratory complications including central and/or obstructive sleep apnoea (estimated at >16% by adolescence)¹⁶. While achondroplasia with co-morbid congenital heart disease is rare, those individuals have twice the risk of heart disease-related death when compared to the general population. In males, it is nearly three times higher, and between ages 25 and 35, heart-disease-related mortality was greater than 10 times higher¹⁷. The pathophysiology of increased heart disease risk among those with achondroplasia is debated but hypotheses include narrower arteries, intrauterine growth retardation, abdominal obesity and hypertriglyceridaemia. Specific data is not available for athletes with achondroplasia.

Visually impaired

The cardiovascular considerations of visually impaired athletes depend on the aetiology of the impairment. Type 1 diabetes resulting in retinopathy underlies the visual impairment in some Paralympic athletes which is also associated with an increased risk of CV disease, with the degree of metabolic control being inversely associated with BP control¹⁸. Acquired visual impairments and inherited disorders such as oculocutaneous and ocular albinism have

not been associated with cardiovascular disease.

CONSIDERATIONS FOR SCREENING

In December 2011, the International Paralympic Committee (IPC) approved the 'Medical Code' which recommends pre-participation evaluations at regular intervals. Although the IPC does not make specific recommendations for the components of the pre-participation evaluation (PPE), it states that "the fitness test should be based on the latest recognised medical knowledge¹⁹."

In comparison, the PPE in AB athletes has evolved over recent years with continued refinement of the criteria used to interpret the resting 12-lead electrocardiogram (ECG) in athletes²⁰⁻²². The 2009 International Olympic Committee (IOC) Consensus Statement on Periodic Health Evaluation of Elite Athletes recommends that "a 12-lead ECG...be recorded on a non-training day, during rest, according to best clinical practice²³." More recently, in 2016, a position statement from the American Medical Society for Sports Medicine (AMSSM) recognises the limitations to the traditional PPE with history and physical exam and the added value of ECG. It also recognises the limited outcomes-based research on the effectiveness of PPEs (with and without ECG) for the prevention of SCD in sport and the lack of infrastructure in many situations for implementing an ECG screening programme. Therefore, the AMSSM position statement recommends an individualised approach for each team considering the available resources and estimated risk of SCD²⁴.

While the debate continues regarding the usefulness of ECG screening in AB athletes, the cost-benefit of screening in Paralympic athletes is more favourable due to the higher prevalence of CV abnormalities associated with sudden death (2% in Paralympic athletes compared to 0.3% in AB athletes)². Additional considerations for thorough CV screening in Paralympic athletes include the increased risk of CHD, metabolic syndrome and potential inability to report CV symptoms, as athletes with neurologic injury such as SCI may have decreased sensory feedback of angina.

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TABLE 1
<i>Parasympathetic responses</i>
<i>Flushing +/- red blotches on face and neck</i>
<i>Bradycardia</i>
<i>Pupillary constriction</i>
<i>Nasal congestion</i>
<i>Sympathetic responses</i>
<i>Profuse sweating (initially above the level of SCI and later below)</i>
<i>Pale, cool, clammy skin below the level of SCI</i>
<i>Piloerection (goose bumps) below the level of SCI</i>
<i>Headache (pounding sensation)</i>
<i>Blurred vision</i>
<i>General apprehension, anxiety</i>

Table 1: Symptoms and Signs of AD.

TABLE 3
<i>1. Sit the athlete upright for potential orthostatic reduction of BP</i>
<i>2. Loosen tight clothing or constrictive devices</i>
<i>3. Check bladder first for distention (catheterize bladder or check for indwelling catheter drainage via collection bag)</i>
<i>4. If symptoms persist and SBP >150 mmHg in adults; >140 in adolescents; or >130 in children 6-12 years old, administer anti-hypertensive agent*</i>
<i>5. If symptoms persist and SBP <150 mmHg, check for bowel impaction using 1% lidocaine jelly and gently disimpact if necessary</i>
<i>6. If symptoms persist despite above measures, scan for other noxious stimuli including new injuries</i>
<i>7. Monitor BP and HR every 5 minutes until resolution</i>

** Topical nitrates to chest can be applied then wiped off once source is found. Nifedipine 10 mg sublingual (chew and swallow) or captopril 25 mg sublingual followed by nifedipine 5 mg if BP remains elevated 30 minutes later.*

Table 3: Treatment algorithm for AD.

TABLE 2
<i>Bladder</i>
<i>Overfilled bag</i>
<i>Cather obstruction</i>
<i>Urinary tract infection</i>
<i>Bowel</i>
<i>Constipation</i>
<i>Fecal impaction</i>
<i>Hemorrhoid</i>
<i>Anal fissure</i>
<i>Skin</i>
<i>Direct irritant (sharp object)</i>
<i>Pressure injuries</i>
<i>Ingrown toenails</i>
<i>Thermal burns/sunburn</i>
<i>Tight clothing</i>
<i>Other</i>
<i>Deep vein thrombosis</i>
<i>Menstrual cramps</i>
<i>Acute abdominal issues</i>
<i>Occult fractures/heterotopic ossification</i>

Table 2: Causes of AD.

Specifically regarding the use of an echocardiogram for screening or as a secondary evaluation, 'grey-zone' values for LV wall thickness may require adjustment in Paralympic athletes with SCI. Since Paralympic hand cyclists with cervical SCI were found to have less extensive concentric left ventricular hypertrophy (LVH), if such an athlete has a LV wall thickness within the currently accepted 'grey-zone', it may be more suggestive of pathologic LVH⁴.

Due to the high incidence of co-morbid cardiovascular disease in athletes with history of stroke, the American Heart Association recommends graded exercise testing with ECG monitoring as part of the PPE⁴.

CONSIDERATIONS FOR EVENT COVERAGE

Orthostatic hypotension

Prevention of orthostasis includes adequate fluid intake, avoidance of diuretics (alcohol and caffeine), elastic compression stockings, abdominal binders and small meals to avoid post-prandial hypotension. In the event of severe symptomatic orthostatic hypotension, the athlete can be placed in a supine or Trendelenburg position and given intravenous (IV) fluids or oral midodrine for blood pressure support. However, in competition, both IV fluids given outside the hospital and midodrine are prohibited by the World Anti-Doping Agency (WADA). If necessary, an athlete may apply for a therapeutic use exemption (TUE) after the fact, if given in an emergency.

Autonomic dysreflexia

For athletes with SCI, the common symptoms of AD reflect the dominance of the parasympathetic nervous system above the level of the SCI and the sympathetic nervous system dominance below the level of the SCI (Table 1). However, even with a significant increase in BP, episodes of autonomic dysreflexia can be asymptomatic in athletes with cervical or high-thoracic injuries.

Any inciting noxious event below the level of SCI can trigger an episode of AD, with bladder and bowel conditions causing 90% of cases of AD (Table 2). With a variable clinical presentation, AD is best treated with an understanding of the presentation by the individual athlete. For example, during athletic training and competition, facial

flushing and sweating above the level of SCI may appear as expected with vigorous exercise. Sweat glands are innervated by the sympathetic nervous system at levels T1-L2, with sweat glands in the upper body from T1-5 and lower part T5-L2. As sympathetic output increases, there is an increase in sweat secretion above the level of injury. However, sweating below the level of injury can also occur as a result of the massive autonomic response that can occur with high-level SCI.

As a potential medical emergency, the initial management is to identify the noxious stimulus to quickly lower the blood pressure and avoid the aforementioned life-threatening complications. The head should be elevated and the legs lowered and any constricting clothing should be loosened. Once symptoms and signs are recognised, the athlete's blood pressure should be checked. As previously described, athletes with SCI above T6 level will have lower baseline resting BP. If the measured BP is 20 to 40 mmHg above the athlete's baseline BP, medical providers should initiate the treatment algorithm (Table 3)²⁵.

After an episode of AD, blood pressure and HR must return to the athlete's normal baseline resting levels and ideally remain there for 2 hours prior to return to sport.

There should be resolution of all symptoms including headaches. To prevent future episodes, all irritants should be removed from the athlete, wheelchair and field of play. If the athlete shows predisposition to autonomic dysreflexia, pharmacologic prophylaxis with alpha-1 adrenoceptor blockers (terazosin 1 to 10 mg adults or 1 to 2 mg children, prazosin 0.5 to 1 mg, 2 or 3 times per day) could be considered, but may only be partially successful for prophylaxis⁹.

Boosting

Boosting is an intentional induction of AD for performance enhancement by competitive wheelchair athletes with cervical and high-thoracic SCI. Due to autonomic dysfunction, athletes with SCI T6 and above may have limited cardiopulmonary exercise capacity with an attenuated response to increase cardiac output and HR. Specifically, these athletes may only achieve a maximal HR between 110 to 130 beats per minute due to impaired sympathetic innervation to the heart.

Presently, there has been no published documentation of an athlete experiencing a dangerous consequence from AD. However, anecdotal accounts of athletes with SCI purposely inducing dysreflexia to enhance performance have been reported.



Studies have shown that boosting the cardiovascular system can improve an athlete's physiologic response to exercise by increasing blood pressure, heart rate and catecholamine release. One study showed 'boosted' wheelchair racers had 10% faster finishing times during a simulated 7.5 km race when compared to 'non-boosted' athletes²⁶.



This method of 'doping' may result in a significant rise in blood pressure before or during competition to compensate for their blunted physiologic response. With goals to 'boost' their attenuated BP and HR responses to exercise, athletes commonly induce AD by intentionally distending their bladders (by consuming large amount of fluids or clamping/kinking their catheters) or excessively tightening waist or leg straps.

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Currently, the International Paralympic Committee (IPC) has banned boosting as a method for performance enhancement, with a penalty of disqualification from Paralympic competitions. In attempts to deter this dangerous method, the IPC has initiated an Operational Management Plan to deter the practice of boosting at major international competitions and with the ultimate goal of protecting athlete health. In one report, it was noted that during the 2008 Beijing Paralympic Games, 2011 ParaPan American Games and 2012 London Paralympic Games, a total of 78 wheelchair track racers and hand cyclists whose functional classification identified their high SCI level were tested²⁸. For the testing protocol, boosting was defined as AD within 2 hours from the start of their competition. A positive test was determined by two sequential recordings of systolic blood pressure >180 mmHg. Since the implementation of the IPC current testing protocol at three international competitions, no athlete has tested positive or has been withdrawn from competition. However, in April 2016, the IPC acknowledged a few limitations of the current protocol and further refined the testing protocol by reducing the threshold systolic blood pressure from 180 mmHg to 160 mmHg for the 2016 Rio Paralympic Games. Results from the refined testing protocol are not yet available.



CONCLUSION

Medical professionals caring for Paralympic athletes must understand the cardiovascular differences that occur as a result of congenital and acquired disability, including physiologic cardiovascular adaptations and structural changes to the myocardium. Differences that are not accounted for may ultimately lead to catastrophic consequences. Sports medicine providers must take these risks into consideration when conducting the periodic health evaluation or providing event coverage for Paralympic sports. The understanding of unique cardiovascular concerns with appropriate management will best support optimal performance in a growing population of athletes with disabilities.

References available at
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