Introduction
Down syndrome (DS) is one of the major causes of mental retardation in this country. One in every 600 individuals born in the United States is affected by this chromosomal anomaly. The phenotypic results of DS can vary greatly. The most frequent findings include mental retardation, orthopedic concerns, cardiac anomalies, vision problems, and epilepsy. In addition, individuals with DS are at greater risk for obesity, and have been shown to have decreased cardiorespiratory capabilities, even when congenital cardiac anomalies are not present.

A physician providing preparticipation care for a person with DS is in the position to assess the nature and extent of motor, sensory, musculoskeletal, mental, and emotional disabilities present that will affect the individual's chances of safely and successfully participating in a sporting activity. Some of these deficits, such as visual refractive errors, may require minor and straightforward adjustments. Other findings, such as atlantoaxial (AA) instability, could prove to be life-threatening, and require strict limitations on certain physical activities.

This article offers the primary care physician a review of the more common and significant conditions found in persons with DS. One goal of this article is to prepare physician readers to consider these conditions as a part of the preparticipation assessment of persons with DS. As a result of this assessment, the physician will be in a better position to provide an exercise prescription, participate in the selection of appropriate sporting events, and assist in developing a better understanding between the patient, parents, coaches, and teachers of the expected outcomes of sport participation by a person with DS.

Medical Conditions Associated with DS
Orthopedic considerations
AA instability is the orthopedic variant in individuals affected by DS that carries the most serious potential concerns in relation to physical activity and sports participation. It has been recommended that persons with DS who have radiographic evidence of AA instability be restricted from participating in sports that entail a high risk of head and neck trauma in order to prevent the occurrence of an acute and catastrophic spinal cord injury [1,2]. Sports in this category include most contact sports such as soccer, football, and basketball, as well as gymnastics, diving, and swimming with a diving start.

In addition, individuals with DS who are scheduled to undergo otolaryngologic surgical procedures or general anesthesia are thought to be at similar risk for spinal cord injury to those persons who wish to participate in high-risk sports [3,4]. The upper airway procedures involve marked extension and rotation of the neck, which may create risk to the spinal cord in individuals with AA instability. During general anesthesia, muscle tone in the cervical region is reduced to the point that previously undetected instability of the AA joint may be unmasked.

The precautions regarding head positioning during general anesthesia in individuals with AA instability do have significance in relation to sport participation. Should cardiopulmonary resuscitation be required for a sports participant with known AA instability, similar care should be taken to avoid excessive jaw thrust or head tilt during resuscitation. These precautions, combined with the common head and neck anomalies present in persons with DS (larger tongue, shorter neck length, and decreased mandible motion), can make cardiopulmonary resuscitation more challenging [5].

Approximately 10% to 40% of all DS patients have been found to exhibit the radiographic findings of an increase in distance between the anterior surface of the odontoid process, and the posterior aspect of the anterior atlas arch [6-8]. This increase in distance signifies...
an excess of ligamentous laxity of the transverse and alar ligaments of the AA joint [9]. The instability cannot be detected by examination or clinical criteria [10]. Factors that have been found to affect the range of diagnosis of this radiographic disorder include the following [9]:

1. The type of radiographic study used (plain films, CT, or MRI)
2. The population studied (adult or pediatric)
3. The specific measurement used by the radiologist to evaluate the degree of instability (measured distance between the bony surfaces of the AA joint and neural canal width are two of the techniques that have been recommended)
4. Whether consideration of bony anomalies of the cervical spine was given in making the diagnosis

Clinical symptoms of AA instability requiring treatment are rare, occurring in 1% to 2% of individuals [11]. These symptoms include neck pain and tilt, gait abnormalities, hypertonicity of the lower extremities, weakness, and bowel or bladder incontinence. These symptoms, associated with cord compression, generally occur before age 10, are more frequent in girls, and are generally chronic rather than acute.

The correlation of the radiographic diagnosis of AA instability with neurologic abnormalities is not well established. A number of studies have shown that the radiographic and clinical findings are poorly related [12], or the radiographic cervical spine findings are clinically insignificant [13–16]. As a result of this poor correlation between radiographic findings and clinical findings, there are no evidence-based guidelines in relation to screening and activity restriction for AA instability. There are several reviews that suggest that individuals with DS should not be screened for AA instability, or barred from high-risk sports and activities [8,17].

The absence of strong evidence in favor of radiographic screening for AA in all individuals with DS is confounded by two significant consensus guidelines in regard to this question. The Special Olympics organization has required routine radiographic screening for AA in all athletes with DS since 1983. The American Academy of Pediatrics confirmed this decision in 1984. In 1995, the AAP recommendation was altered, acknowledging the lack of evidence in support of routine radiographic screening for AA instability. However, the panel did not change the existing recommendation for screening. Because of these guidelines and the life-changing, as well as medical-legal, consequences of the rare acute neurospinal injury in individuals with DS, consideration must be given to some type of screening for AA instability until there are new guidelines based on controlled prospective trials.

The decision in regard to if, when, and how to screen for AA instability should be made by the parents or legally designated caregivers of an individual with DS after they have had the opportunity to consider the potential benefits and limits of screening. Important points to include in this discussion are as follows [9]:

1. Screening is recommended for those persons with DS who wish to participate in high-risk physical activities (or those who are going to have head and neck surgery, or undergo general anesthesia)
2. Screening recommendations include a plain film of the cervical spine, combined with an MRI of the same region
3. Individuals with bony anomalies of the AA or the occipitoatlantal junction are at high risk, regardless of MRI findings, and should be referred to a pediatric spine surgeon
4. Asymptomatic individuals with a normal atlantodens interval (ADI) on plain films, and a neural canal width (NCW) of > 14 mm are normal; because these radiographic findings have not been found to change over time [16,18,19], repeat radiographic studies are not indicated
5. Individuals with an abnormal ADI (> 4.5 mm) or an NCW of < 14 mm would also benefit from consultation with a pediatric spine surgeon

Additional orthopedic findings that may affect sports participation, and that are more frequently found in individuals with DS include patellar-femoral instability, valgus and varus deformities of the halux, pes planus, scoliosis, and general decrease in muscle tone.

These problems are thought to be related to the generally increased ligamentous laxity of individuals with DS. Knowledge of the increased presence of these musculoskeletal problems may allow a treating physician to provide more directed assessment, treatment, and preventive care to reduce the possibility of a sports related injury or overuse syndrome. Accommodation and treatment of these problems may include a number of approaches, such as guidance on activity selection and limitations, directed physical therapy to build muscle strength for joint stability, and the use of supportive taping and orthotics.

Cardiovascular considerations in DS
Cardiac anomalies are found in 40% to 50% of persons with DS. In addition, individuals with DS with and without cardiac malformations have been found to have lower cardiovascular fitness levels than their peers [20]. These findings, combined with upper airway malformations of