Complete Summary

GUIDELINE TITLE

Cardiovascular health supervision for individuals affected by Duchenne or Becker muscular dystrophy.

BIBLIOGRAPHIC SOURCE(S)

American Academy of Pediatrics Section on Cardiology and Cardiac Surgery. Cardiovascular health supervision for individuals affected by Duchenne or Becker muscular dystrophy. Pediatrics 2005 Dec;116(6):1569-73. [60 references] PubMed

GUIDELINE STATUS

This is the current release of the guideline.

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COMPLETE SUMMARY CONTENT

SCOPE

DISEASE/CONDITION(S)

- Duchenne muscular dystrophy
- Becker muscular dystrophy
- Dilated cardiomyopathy
- Cardiac arrhythmia

GUIDELINE CATEGORY

Evaluation
Management
Risk Assessment
Screening
Treatment

CLINICAL SPECIALTY

Cardiology
Family Practice
Internal Medicine
Pediatrics

INTENDED USERS

Physicians

GUIDELINE OBJECTIVE(S)

To provide recommendations for optimal cardiovascular evaluation to health care specialists caring for individuals with Duchenne or Becker muscular dystrophy

TARGET POPULATION

Individuals with Duchenne or Becker muscular dystrophy

INTERVENTIONS AND PRACTICES CONSIDERED

1. Complete cardiac evaluation, including:
    - History and physical examination
    - Electrocardiogram
    - Transthoracic echocardiogram
    - Multigated acquisition study (MUGA) or cardiac magnetic resonance imaging (MRI) for patients with limited echocardiographic acoustic windows
2. Pharmacologic treatment of cardiac dysfunction (e.g., diuretics, angiotensin-converting enzyme inhibitors, beta-blockers); rhythm abnormalities; and respiratory abnormalities
3. Increased cardiovascular surveillance for patients undergoing treatment with glucocorticoids
4. Pre-operative evaluation of baseline cardiac and pulmonary function
5. Intraoperative cardiac monitoring
6. Postoperative maintenance of fluid balance and cardiopulmonary monitoring
7. Optimization of nutrition status
8. Anticoagulation therapy
9. Optimizing pulmonary care
10. Cardiovascular screening of female carriers of Duchenne or Becker muscular dystrophy

MAJOR OUTCOMES CONSIDERED

Morbidity and mortality associated with cardiovascular complications of Duchenne and Becker muscular dystrophy

METHODOLOGY

METHODS USED TO COLLECT/SELECT EVIDENCE

Searches of Electronic Databases

DESCRIPTION OF METHODS USED TO COLLECT/SELECT THE EVIDENCE

Not stated

NUMBER OF SOURCE DOCUMENTS

Not stated

METHODS USED TO ASSESS THE QUALITY AND STRENGTH OF THE EVIDENCE

Not stated

RATING SCHEME FOR THE STRENGTH OF THE EVIDENCE

Not applicable

METHODS USED TO ANALYZE THE EVIDENCE

Review

DESCRIPTION OF THE METHODS USED TO ANALYZE THE EVIDENCE

Not stated

METHODS USED TO FORMULATE THE RECOMMENDATIONS

Expert Consensus

DESCRIPTION OF METHODS USED TO FORMULATE THE RECOMMENDATIONS

Not stated

RATING SCHEME FOR THE STRENGTH OF THE RECOMMENDATIONS

Not applicable

COST ANALYSIS

A formal cost analysis was not performed and published cost analyses were not reviewed.

METHOD OF GUIDELINE VALIDATION

Peer Review

DESCRIPTION OF METHOD OF GUIDELINE VALIDATION

Not stated

RECOMMENDATIONS

MAJOR RECOMMENDATIONS

Recommendations for Cardiac Care in Patients With Duchenne Muscular Dystrophy (DMD) or Becker Muscular Dystrophy (BMD)

1. Cardiac care of the patient with DMD or BMD should begin after confirmation of the diagnosis. The patient should be referred for evaluation to a cardiac specialist with an interest in the management of cardiac dysfunction and/or neuromuscular disorders.
2. A complete cardiac evaluation should include (but not be limited to) a history and physical examination, electrocardiogram, and transthoracic echocardiogram. Consideration should be given to a multigated acquisition study (MUGA) or cardiac magnetic resonance imaging (MRI) in patients with limited echocardiographic acoustic windows.
3. Clinicians should be aware that the typical signs and symptoms of cardiac dysfunction may not be present secondary to the patient's musculoskeletal limitations. Weight loss, cough, nausea and vomiting, orthopnea, and increased fatigue with a decreased ability to tolerate the daily regimen may represent cardiac impairment and should be investigated. However, the development of dilated cardiomyopathy usually precedes the development of heart-failure symptoms by years and must be identified at its earliest onset.
4. Signs and symptoms of cardiac dysfunction should be treated. Consideration should be given to the use of diuretics, angiotensin-converting enzyme inhibitors, and/or beta-blockers.
5. Abnormalities of cardiac rhythm should be promptly investigated and treated. Periodic Holter monitoring should be considered for patients with demonstrated cardiac dysfunction.
6. Respiratory abnormalities contribute to the cardiovascular morbidity and mortality of the disease. Concurrent evaluation and treatment of respiratory abnormalities are recommended.
7. Individuals undergoing treatment with glucocorticoids warrant increased cardiac surveillance with specific monitoring for weight gain and hypertension.
8. Complete cardiac evaluation should be undertaken before scoliosis surgery or other major surgical procedures. Consideration should be given to cardiac stress testing (such as a dobutamine stress echocardiogram) if abnormalities of cardiac function are present during resting evaluation. Medical therapy should be optimized before surgery, and the risks and benefits of the procedure should be discussed in detail with the patient and the family.

9. Intraoperative cardiac monitoring should be undertaken in individuals with DMD or BMD during major surgical procedures. Specific anesthetic techniques and decisions about intraoperative ventilation will depend on the patient and the procedure. Agents known to trigger hyperkalemia (e.g., succinylcholine chloride) or a hypermetabolic state (e.g., inhaled anesthetic agents) should be avoided. Cardiac monitoring should continue in the postoperative period.
10. Anticoagulation therapy should be considered in patients with severe cardiac dysfunction to prevent systemic thromboembolic events.
11. Clinicians who are experienced in the care of patients with DMD or BMD and are knowledgeable about the pathogenesis of the disease should be actively involved when patients are treated in an intensive care setting.
12. Nutritional status should be optimized to the special needs of patients with DMD or BMD.

Recommendations Specific for Cardiac Care in Patients with DMD

1. Patients should be routinely managed in early childhood with a complete cardiac evaluation at least biannually.
2. Yearly complete cardiac evaluations should begin at approximately 10 years of age or at the onset of cardiac signs and symptoms. However, individuals demonstrating these signs and symptoms are relatively late in their course.

Recommendations Specific for Cardiac Care in Patients with BMD

1. Complete cardiac evaluations should begin at approximately 10 years of age or at the onset of signs and symptoms. Evaluations should continue at least biannually.

Recommendations for Cardiac Care in Carriers of DMD or BMD

1. Carriers of DMD or BMD should be made aware of the risk of developing cardiomyopathy and educated about the signs and symptoms of heart failure.
2. Carriers of DMD or BMD should be referred for evaluation by a cardiac specialist with experience in the treatment of heart failure and/or neuromuscular disorders. Patients should undergo initial complete cardiac evaluation in late adolescence or early adulthood or at the onset of cardiac signs and symptoms, if these signs or symptoms appear earlier.
3. Carriers should be screened with a complete cardiac evaluation at a minimum of every 5 years starting at 25 to 30 years of age.
4. Treatment of cardiac disease is similar to that outlined for boys with DMD or BMD.

CLINICAL ALGORITHM(S)

None provided

EVIDENCE SUPPORTING THE RECOMMENDATIONS

TYPE OF EVIDENCE SUPPORTING THE RECOMMENDATIONS

The type of evidence supporting the recommendations is not specifically stated.

BENEFITS/HARMS OF IMPLEMENTING THE GUIDELINE RECOMMENDATIONS

POTENTIAL BENEFITS

- Appropriate cardiovascular monitoring and evaluation of patients with Duchenne or Becker muscular dystrophy may lead to decreased morbidity and mortality in these patients.
- Investigation of cardiomyopathy in patients with Duchenne muscular dystrophy and Becker muscular dystrophy, as well as carriers, will benefit affected individuals, and new knowledge may lead to the elucidation of novel treatment strategies for dilated cardiomyopathy.

POTENTIAL HARMS

Not stated

IMPLEMENTATION OF THE GUIDELINE

DESCRIPTION OF IMPLEMENTATION STRATEGY

An implementation strategy was not provided.

INSTITUTE OF MEDICINE (IOM) NATIONAL HEALTHCARE QUALITY REPORT CATEGORIES

IOM CARE NEED

Living with Illness

IOM DOMAIN

Effectiveness

IDENTIFYING INFORMATION AND AVAILABILITY

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ADAPTATION

Not applicable: The guideline was not adapted from another source.

DATE RELEASED

2005 Dec

GUIDELINE DEVELOPER(S)

American Academy of Pediatrics - Medical Specialty Society

SOURCE(S) OF FUNDING

American Academy of Pediatrics

GUIDELINE COMMITTEE

Section on Cardiology and Cardiac Surgery

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FINANCIAL DISCLOSURES/CONFLICTS OF INTEREST

Not stated

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GUIDELINE AVAILABILITY

Electronic copies: Available from the American Academy of Pediatrics (AAP) Policy Web site.

Print copies: Available from American Academy of Pediatrics, 141 Northwest Point Blvd., P.O. Box 927, Elk Grove Village, IL 60009-0927.

AVAILABILITY OF COMPANION DOCUMENTS

None available

PATIENT RESOURCES

None available

NGC STATUS

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