Health Policy & Clinical Effectiveness Program

Evidence-Based Care Guideline

Therapy Management of Congenital Muscular Torticollis

in children age 0 to 36 months

Publication Date: 03-17-09

Target Population

Inclusions: These guidelines are intended for use in patients 0 to 36 months of age with a diagnosis of congenital muscular torticollis (CMT) and demonstrated cervical lateral flexion and/or rotation limitations of greater than 5º.

Exclusions: These guidelines are not intended for use with patients with the following:

- demonstrated cervical lateral flexion and/or rotation limitations of less than 5º
- sudden onset torticollis
- posterior fossa tumor
- hemiplegia
- abnormal vertebral structure (e.g. Klippel-Feil Syndrome, hemivertebrae)
- abnormal alignment of the cervical vertebrae
- fracture and/or dislocation of any vertebrae
- other conditions that would contraindicate passive movement of the cervical spine

Target Users

Included but are not limited to:

- Physical Therapists
- Occupational Therapists
- patients and families/caregivers
- primary care providers

Introduction

References in parentheses ( ) Evidence strengths in [ ] (See last page for definitions)

Torticollis literally means "twisted neck or "wryneck" and is derived from the Latin words "torquere" (twisted) and "collum" (neck) *(Stellwagen 2004 [S])*. The generic term "torticollis" is not a specific diagnosis but a word used to describe the twisted neck posture *(Scoles 1988 [E])*. It may be a clinical sign of one of a variety of underlying pathologies, some benign and some quite serious *(Cheng 2000 [C], Ballock 1996 [D])*.

The torticollis posture in an infant or child may result from a muscular, skeletal, neurological, inflammatory or neoplastic condition. Determining the origin of the torticollis is critical to an accurate diagnosis and appropriate intervention. Over 80% of all infants presenting with a torticollis posture will be found to have congenital muscular torticollis (CMT). In the remaining 20%, the torticollis posture may represent a sign of a more serious underlying condition. Therefore it is critical when confirming a diagnosis of CMT that a muscular origin be identified.

CMT is a musculoskeletal condition observed at birth or early infancy resulting from unilateral fibrosis and shortening of the sternocleidomastoid muscle (SCM) *(Karmel-Ross 2006 [E])*. The diagnosis of CMT is based upon a careful perinatal history and the examination and may be confirmed by ultrasonography of the involved SCM *(Cheng 2000 [C])*. Typically, the infant presents with limitation of active and passive neck motion, a posturing of the neck in lateral flexion to the ipsilateral side and rotation to the contralateral side causing the chin to point toward the contralateral shoulder. CMT is the third most frequently occurring musculoskeletal condition in infants with a reported incidence of 0.4% to 1.9%. CMT is often seen in combination with metatarsus adductus and developmental hip dysplasia. All three congenital deformities are associated with fetal intrauterine malposition, although the exact etiology of CMT appears to be multifactorial and remains uncertain *(Cooperman 1997 [S])*.

Eighty percent of torticollis in infancy is determined to be muscular in origin and correctly termed CMT *(Ballock 1996 [D])*. Infants with CMT may be classified into one of three groups based on initial clinical presentation:

1. Sternomastoid tumor group (SMT);
2. Muscular torticollis without palpable tumor group (MT);
3. Postural torticollis group (POST), clinical features of CMT without sternomastoid tumor or muscle tightness *(Cheng 2000 [C])*.

The best predictors of successful outcome are clinical group, severity of deficit of neck rotation and age at initiation of therapy *(Cheng 2001 [C])*.

Over 80% of infants with CMT also present with craniofacial asymmetry and deformational plagiocephaly (DP) of varying degree *(Cheng 2000 [C], Cheng 1994 [D])*;

and one study reports that 20% of infants referred for evaluation of DP also had torticollis *(Cheng 1994 [D])*. DP, also referred to as posterior positional plagiocephaly or plagiocephaly without stenosis, describes an asymmetry of the infant skull or face not resulting from stenosis or premature closure of the cranial sutures. Physical therapy interventions for CMT may also improve positional preference and DP *(van Vlimmeren 2008 [A], Freed 2004 [S])*.

Up until the mid 1960's, the standard treatment for CMT was surgical release of the sternocleidomastoid muscle (SCM) during the first few months of life and was followed by a lengthy period of immobilization in an overcorrected position. Over the next 25 years clinical studies revealed that good outcomes could be achieved for CMT when treated conservatively with physical therapy and without surgery.

In 1992 the American Academy of Pediatrics formally introduced the recommendation that all infants be positioned in supine for sleeping with a "Back to Sleep" campaign *(AAP 1992 [E])*. The following decade saw a dramatic increase, indeed an "epidemic" *(de Chalain 2005 [D])* in the number of infants referred to physical therapy for treatment of CMT and the associated DP *(Freed 2004 [S])*.

The objectives of this guideline are to:

- promote appropriate referrals
- provide optimal skilled care to patients
- decrease unwarranted variation in care
- improve functional outcomes
- improve patient/family satisfaction

Intervention

No higher-level evidence was identified, although a body of moderately strong evidence supports the effectiveness of conservative treatment of CMT *(Cheng 2000 [C], Emery 1994 [D], Binder 1987 [D])*.

Over 90% of children achieve a good to excellent outcome with conservative treatment when therapy is initiated during the first 12 months of life *(Cheng 2000 [C])*. Children in the SCM tumor group, those with an initial deficit in cervical rotation of >30º, and those who initiate therapy after 12 months are more likely to require surgical intervention to attain a functional and cosmetically acceptable outcome *(Cheng 2000 [C])*. Conservative treatment for CMT includes physical therapy and physician observation. Physical therapy interventions include positioning, environmental adaptations, passive and active stretching of the tight SCM, strengthening of weak neck and trunk muscles and movement therapy *(Cheng 2000 [C], Emery 1994 [D], Binder 1987 [D], Freed 2004 [S])*. Cheng et al. advocates a program of manual stretching provided by physical therapists *(Cheng 2001 [C], Cheng 2000 [C])*. In addition, it is recommended that a therapist train the child's caregiver to implement a consistent therapy program at home *(Emery 1994 [D], Binder 1987 [D], Freed 2004 [S])*.

Investigating factors associated with length of treatment duration, Emery reported that when physical therapy was initiated before age 2 years, the average duration of treatment was 4.7 months (SD 5.6 months), although the range varied widely, from 1 to 36 months, among the 100 children that achieved resolution without surgery *(Emery 1994 [D])*.

Note 1: Some children may not achieve optimal outcome with conservative management and may require surgical intervention to achieve optimal functional outcome *(Cheng 2001 [C], Cheng 2000 [C])*.
Note 2: Recent reports describe the use of Botulinum Toxin for some children who do not respond adequately to conservative treatment *(Joyce 2005 [D], Oleszek 2005 [D], Do 2006 [E])*.

Expected Outcome

The majority (>90%) of infants and children diagnosed with CMT achieve good to excellent outcome with conservative treatment *(Cheng 2000 [C], Taylor 1997 [D])*. Conservative treatment may include passive stretching, active movement, strengthening, and positioning *(Freed 2004 [S])*.

Guideline Recommendations

Assessment (see Algorithm 1)

1. It is recommended that a Physical Therapy Examination be completed, including the components named in the Table *(Karmel-Ross 1997 [E])*.

Table: Physical Therapy Examination Components

History	
Parent Report of History	• birth/delivery o length of infant at birth o presence of skull asymmetry at birth o presence of facial asymmetry at birth
Current Health	• feeding • positioning devices used
Torticollis/Plagiocephaly History	• onset • changes in symptoms
Systems Review	
Visual Function	• midline visual focus • ocular alignment
Hip Screen	• asymmetry • hip clunk • leg length discrepancy
Neurological Screen	• predominant Asymmetrical Tonic Neck Reflex (ATNR) • abnormal muscle tone • presence of sustained clonus
Pain Assessment	• appropriate pain scale
Skin Screen	• clinical appearance of neck
Physical Assessment	
Clinical Observations	• resting posture • motor development
Plagiocephaly/ Anthropometrics	• cranial shape • type
Range of Motion	• cervical • upper and lower extremity
Palpation *(Karmel-Ross 1997 [E]), (Kendall 1993 [E])*	• SCM • Trapezius • Scalenes

2. It is recommended that following the assessment, if the patient does not meet the inclusion criteria or meets exclusion criteria for the CMT guideline, that the evaluating therapist immediately contact the referring physician for further consultation regarding appropriate treatment or the need for further medical evaluation *(Ballock 1996 [D])*.
3. It is recommended that an immediate recommendation for referral to the appropriate specialist be made to the referring physician if
   - visual dysfunction is observed (Ophthalmology) *(Ballock 1996 [D])*
   - hip screen is failed (Orthopaedics) *(Cheng 2000 [C])*
   - neurological screen is failed (Neurology) *(Ballock 1996 [D])*
   - patient demonstrates Plagiocephaly Type I through V (Plastic Surgery) *(Argenta 2004 [E])*
   - patient presents with boney end feel (Orthopaedics) *(Ballock 1996 [D])*
   - patient is 18 months or older at presentation *(Local Consensus [E])*.

Treatment Recommendations (see Algorithm 2)

Initial Visit

4. It is recommended that the treatment of CMT begin at the time of the initial therapy evaluation and include:
   - Instruction in home program to include stretching exercises and range of motion exercises (cervical lateral flexion and rotation) *(Freed 2004 [S])*
   - Education on basic pathology of CMT *(Local Consensus [E])*
   - Education on environmental adaptations and appropriate positioning to reduce deformational forces to face and skull *(Freed 2004 [S])*

Second Visit

5. It is recommended that the second visit occur within 2 weeks of the initial examination *(Local Consensus [E])*.
6. It is recommended that the second visit include the components *(Freed 2004 [S])*:
   - review of established home exercise program (parent demonstration)
   - provision of parental instruction on combination stretches, parent hold and strengthening activities
   - re-check of cervical range of motion, skull/facial asymmetry and posture/positioning
   - developmental screen
   - screening of upper extremity and lower extremity range of motion

Subsequent Visits

7. It is recommended that all subsequent visits include:
   - reassessment of range of motion (ROM), strength, and developmental progression
   - review of the Home Education Program (HEP)
   - addition of exercises, developmental activities and environmental adaptations as appropriate

   (Local Consensus [E])

Frequency and Progression of Intervention

8. It is recommended that children age 0 to 4 months at the onset of treatment, be followed every other week to monitor and modify the program as appropriate *(Local Consensus [E])*.

9. It is recommended that children age 4 to 5 months at the onset of treatment, be followed weekly to monitor and to modify the program as appropriate. A higher frequency is recommended when the child enters treatment at this age, due to the child's developmental ability to add strengthening components.
   Home program to include:
   - righting and equilibrium reactions of head and trunk
   - transitional movements and weight shifting
   - upper extremity (UE) weight bearing
   - concentric and eccentric strengthening of head and trunk muscles

   (Freed 2004 [S], Local Consensus [E]).

   Note: Children greater than 4 months of age who demonstrate >10 º of head tilt after 2 to 3 months of treatment, may be assessed for a potential trial of kinesiotaping *(Local Consensus [E])* and/or an orthosis (i.e., collar for tubular orthosis for torticollis [TOT], or a soft orthosis). *(Cottrill-Mosterman 1987 [O], Karmel-Ross 2006 [E], Local Consensus [E])*.

10. It is recommended that children age 6 to 8 months at the onset of treatment, be followed weekly to monitor and to modify the home program as appropriate.
    Home program to include:
    - righting and equilibrium reactions in sitting
    - antigravity strengthening of neck and trunk

    (Freed 2004 [S], Local Consensus [E]).

11. It is recommended that children age 9 to 12 months old at the onset of treatment, be followed weekly to monitor and to modify the program as appropriate.
    Home program to include:
    - age appropriate balance and developmental activities
    - equilibrium reactions in quadruped

    (Freed 2004 [S], Local Consensus [E]).

12. It is recommended that children age 12 to 18 months at the onset of treatment, be followed every 1 to 2 weeks:
    - if head tilt is >5° after child becomes independently ambulatory communicate with the primary care provider to recommend consideration of additional followup by orthopaedics and ophthalmology

    (Local Consensus [E]).

13. It is recommended that children age 18 to 36 months at the onset of treatment be followed every 1 to 2 weeks while awaiting specialty consults:
    - regardless of head tilt or range of motion, communicate with primary care provider to recommend consideration of additional followup by orthopaedics and ophthalmology

    (Local Consensus [E]).

14. It is recommended that the child with residual limitations after 6 months of treatment be referred to the primary care provider for consideration of additional followup:

    Note 1: Continue plan of care while awaiting specialty consults *(Local Consensus [E])*.

    Note 2: Consider a limited trial of soft tissue techniques while awaiting specialty consults *(Local Consensus [E])*.
    - Surgical consult for children with less than 75 degrees of cervical rotation or persistence of palpable tumor *(Cheng 2001 [C], Cheng 2000 [C])*.

      Note: Cheng states that surgical intervention is indicated in the above circumstance *(Cheng 2001 [C])*.
    - Ophthalmology consult for children with residual head tilt with adequate ROM and strength *(Local Consensus [E])*.

Discharge from Therapy

15. It is recommended that a child be discharged from therapy when therapy goals have been achieved. Goals include:
    - cervical range of motion within 5 degrees of normal limits for passive and active lateral flexion and rotation.
    - symmetrical posture in all functional positions.
    - head in midline during activity the majority of the time.
    - symmetrical gross motor skills.

    (Local Consensus [E])

16. It is recommended that parents be instructed that plateaus in active range of motion and /or a temporary decrease in midline head control may occur during times of:
    - gross motor progression, such as attainment of independent ambulation
    - growth spurt
    - ear infection or other illness

At discharge, parents are instructed to resume home exercises during these instances. If the above persists for more than 10 to 14 days, a therapy reassessment may be indicated *(Karmel-Ross 1997 [E])*.

17. It is recommended that after 12 months of treatment, the child is re-evaluated to assess progress toward goals and to determine if there will be a continued benefit from further therapy services. The child may require additional medical intervention to achieve a more functional outcome:
    - communicate with primary care provider to determine future plan of care.

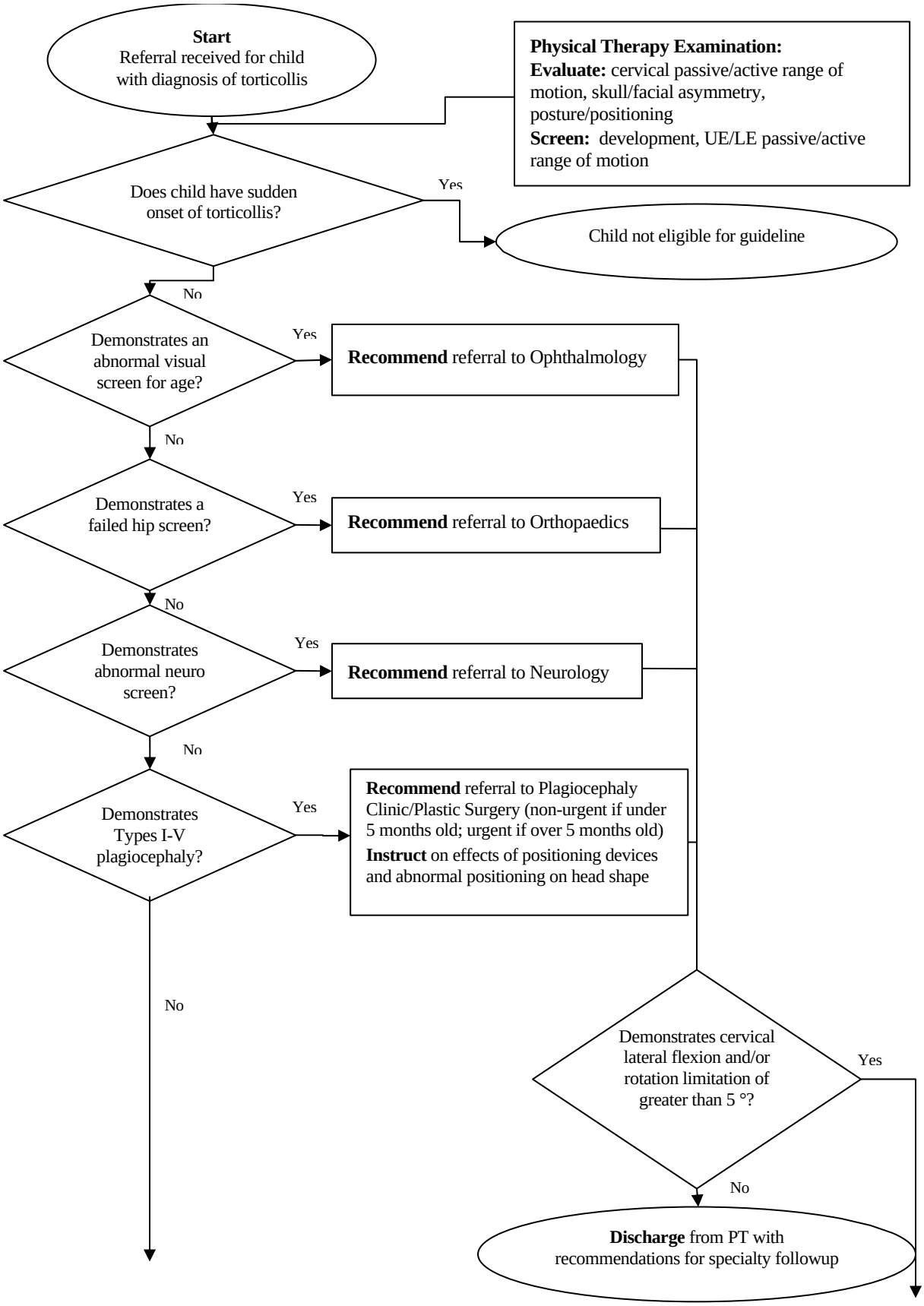
    (Local Consensus [E]).

    Note 1: Additional patient factors may be considered when determining discharge.
    Note 2: Average duration of treatment is reported to be less than 6 months, ranging from 1 to 36 months. Initial severity of the deficit in cervical rotation, a greater head tilt, or presence of a mass predict that a longer treatment duration may be required to achieve optimal outcome *(Emery 1994 [D])*.

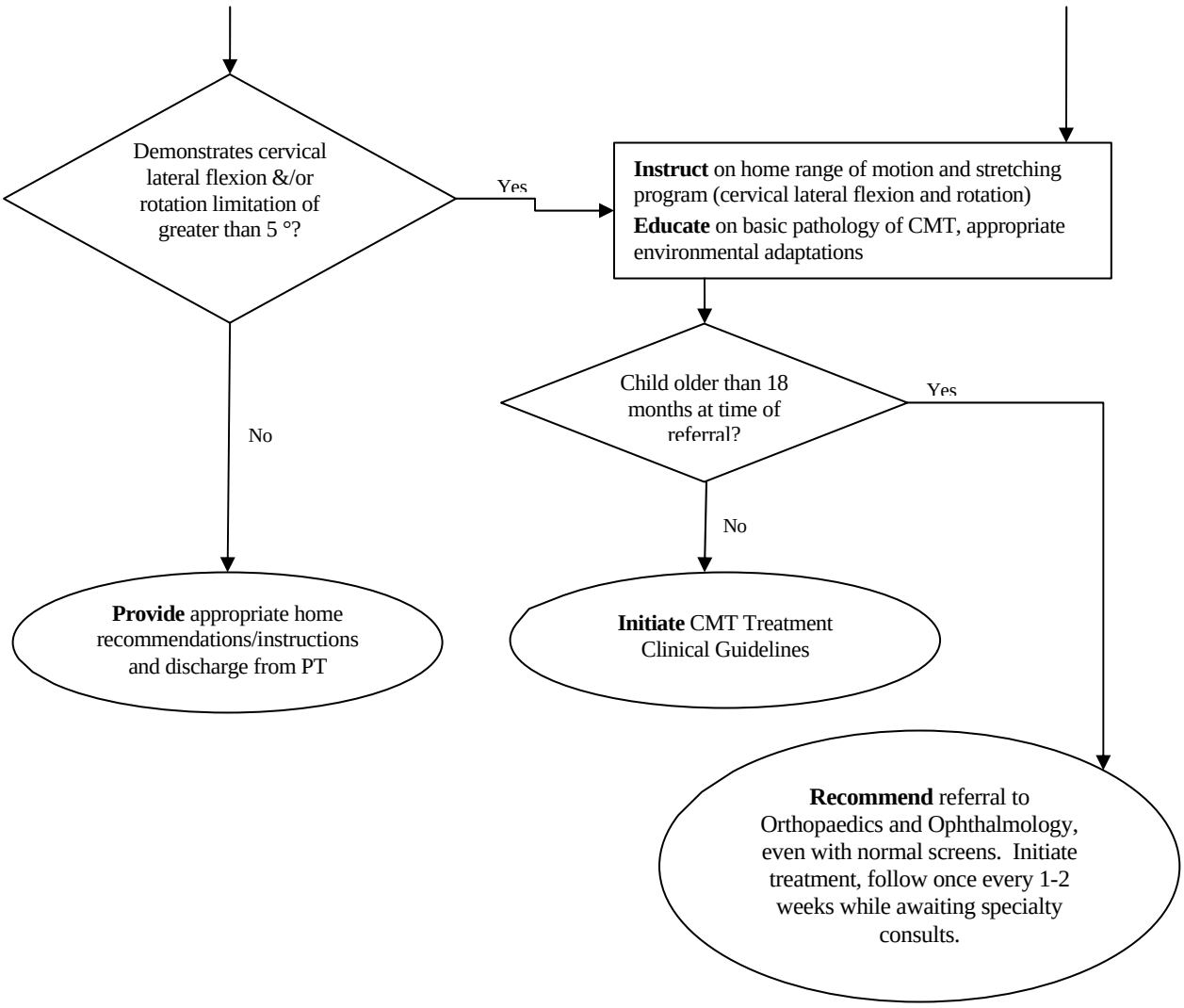
Future Research Agenda

1. Clinical trial evaluating the effectiveness of program of passive stretching compared to a program of active range of motion and positioning (without passive stretching).
2. Clinical trial to determine the frequency of therapy intervention that results in optimal outcome.
3. Clinical trial that more accurately predict which infants will not achieve optimal outcomes with conservative care.
4. Clinical trial evaluating the effectiveness of the TOT collar.

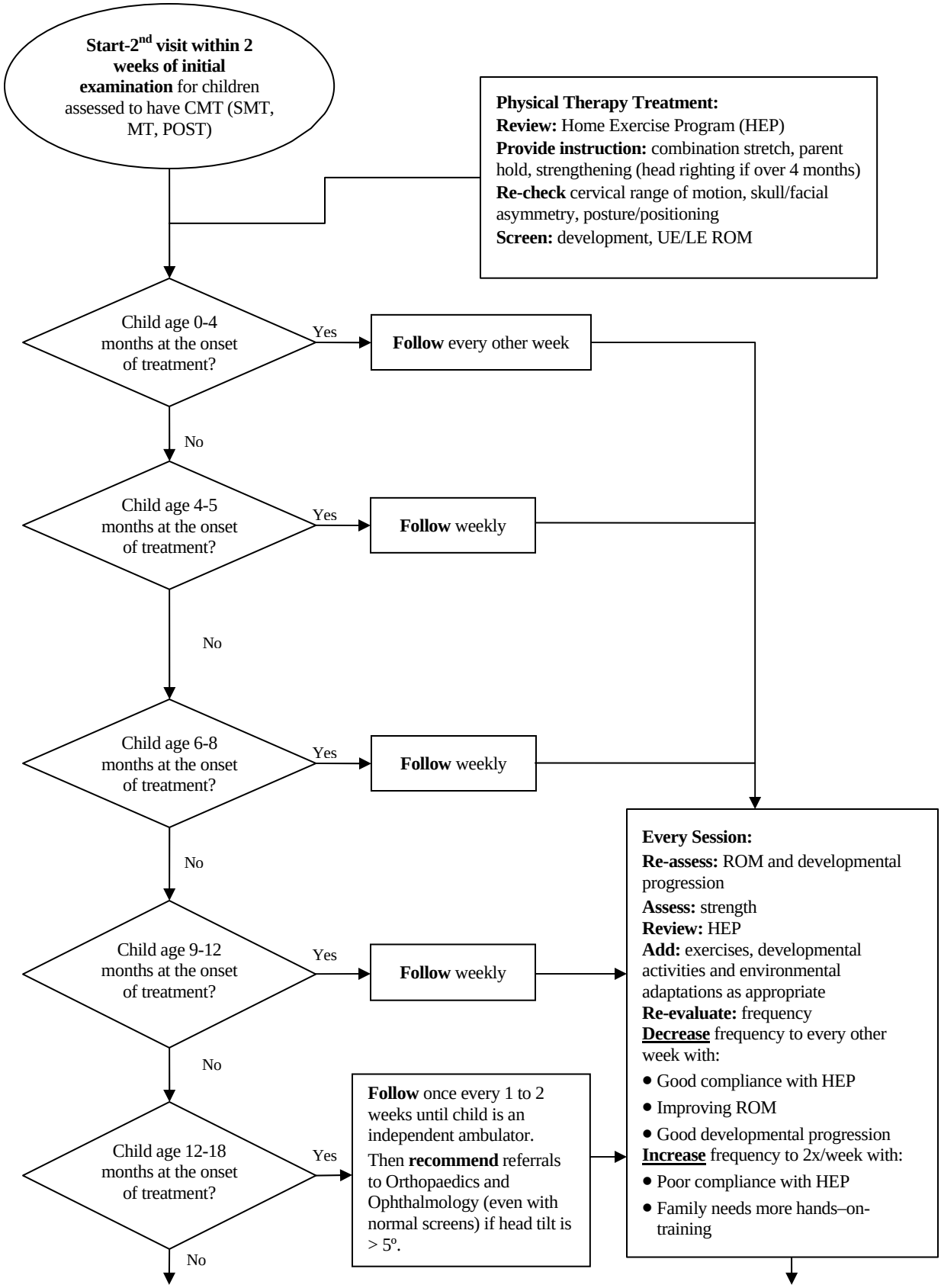
Algorithm 1: CMT Patient Evaluation



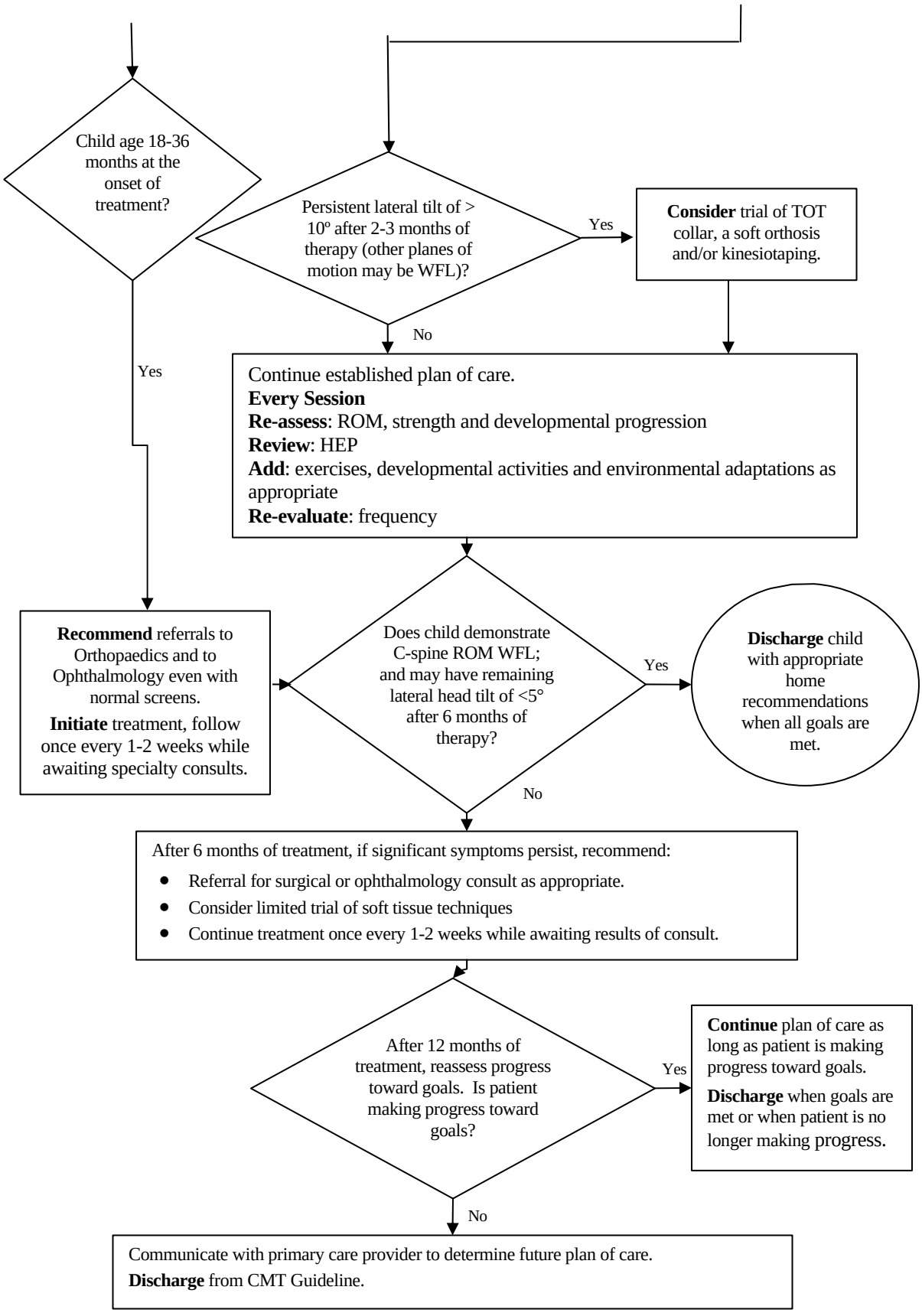
Algorithm 1: CMT Patient Evaluation (continued)



Algorithm 2: CMT Patient Treatment



Algorithm 2: CMT Patient Treatment (continued)



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All Team Members and Clinical Effectiveness support staff listed above have signed a conflict of interest declaration.

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Development Process

The process by which this guideline was developed is documented in the Guideline Development Process Manual; a Team Binder maintains minutes and other relevant development materials. The recommendations contained in this guideline were formulated by an interdisciplinary working group which performed systematic and critical literature reviews, using the grading scale that follows, and examined current local clinical practices.

To select evidence for critical appraisal by the group for the update of this guideline, the Medline, EmBase and the Cochrane databases were searched for dates of January 1965 to April 2008 to generate an unrefined, "combined evidence" database using a search strategy focused on answering clinical questions relevant to congenital muscular torticollis and employing a combination of Boolean searching on human-indexed thesaurus terms (MeSH headings using an OVID Medline interface) and "natural language" searching on searching on human-indexed thesaurus terms (MeSH headings using an OVID Medline interface) and "natural language" searching on words in the title, abstract, and indexing terms. The citations were reduced by eliminating duplicates, review articles, non-English articles, and adult articles. During the course of the guideline development, additional clinical questions were generated and subjected to the search process, and some relevant review articles were identified.

Tools to assist in the effective dissemination and implementation of the guideline may be available online at http://www.cincinnatichildrens.org/svc/alpha/h/health-policy/ev-based/default.htm . Once the guideline has been in place for three years, the development team reconvenes to explore the continued validity of the guideline. This phase can be initiated at any point that evidence indicates a critical change is needed.

CCHMC Grading Scale			
M	Meta-analysis or Systematic Review	O	Other evidence
A	Randomized controlled trial: large sample	E	Expert opinion or consensus
B	Randomized controlled trial: small sample	F	Basic Laboratory Research
C	Prospective trial or large case series	L	Legal requirement
D	Retrospective analysis	Q	Decision analysis
S	Review article	X	No evidence

Recommendations have been formulated by a consensus process directed by best evidence, patient and family preference and clinical expertise. During formulation of these recommendations, the team members have remained cognizant of controversies and disagreements over the management of these patients. They have tried to resolve controversial issues by consensus where possible and, when not possible, to offer optional approaches to care in the form of information that includes best supporting evidence of efficacy for alternative choices.

The guideline has been reviewed and approved by clinical experts not involved in the development process, distributed to senior management of Cincinnati Children's Hospital Medical Center (CCHMC), and other parties as appropriate to their intended purposes.

The guideline was developed without external funding. All Team Members and Clinical Effectiveness support staff listed have declared whether they have any conflict of interest and none were identified.

Copies of this Evidence-based Care Guideline (EBCG) and its any available implementation tools are available online and may be distributed by any organization for the global purpose of improving child health outcomes. Website address: http://www.cincinnatichildrens.org/svc/alpha/h/health-policy/ev-based/default.htm Examples of approved uses of the EBCG include the following:

- copies may be provided to anyone involved in the organization's process for developing and implementing evidence based care guidelines;
- hyperlinks to the CCHMC website may be placed on the organization's website;
- the EBCG may be adopted or adapted for use within the organization, provided that CCHMC receives appropriate attribution on all written or electronic documents; and
- copies may be provided to patients and the clinicians who manage their care.

Notification of CCHMC at HPCEInfo@cchmc.org for any EBCG, or its companion documents, adopted, adapted, implemented or hyperlinked by the organization is appreciated.

NOTE: These recommendations result from review of literature and practices current at the time of their formulations. This guideline does not preclude using care modalities proven efficacious in studies published subsequent to the current revision of this document. This document is not intended to impose standards of care preventing selective variances from the recommendations to meet the specific and unique requirements

of individual patients. Adherence to this guideline is voluntary. The physician in light of the individual circumstances presented by the patient must make the ultimate judgment regarding the priority of any specific procedure.

For more information about this guideline, its supporting evidences and the guideline development process, contact the Division of Occupational Therapy and Physical Therapy Office at 513-636-4651.

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Note: When using the electronic version of this document, "☞" refers to journal articles that have a hyperlink to the abstract. A hyperlink following this symbol goes to the article PDF when the user is within the CCHMC network.

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