



Effective Health Care Program

Interventions for Feeding and Nutrition in Cerebral Palsy

Executive Summary

Background

Cerebral palsy (CP) is a “group of disorders of the development of movement and posture, causing activity limitation, that is attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behaviour, and/or by a seizure disorder.”¹ This group of syndromes ranges in severity and is the result of a variety of etiologies occurring in the prenatal, perinatal, or postnatal period. Though the disorder is nonprogressive, the clinical manifestations may change over time as the brain develops, with other neurologic impairments frequently co-occurring.^{1,2}

More than 100,000 children are estimated to be affected with CP in the United States. Due to advances in supportive medical care, approximately 90 percent of children with CP survive into adulthood, resulting in an additional estimated 400,000 adults living with CP in the United States.³⁻⁸ Lifetime costs are estimated to be nearly \$1 million per person.⁹

Effective Health Care Program

The Effective Health Care Program was initiated in 2005 to provide valid evidence about the comparative effectiveness of different medical interventions. The object is to help consumers, health care providers, and others in making informed choices among treatment alternatives. Through its Comparative Effectiveness Reviews, the program supports systematic appraisals of existing scientific evidence regarding treatments for high-priority health conditions. It also promotes and generates new scientific evidence by identifying gaps in existing scientific evidence and supporting new research. The program puts special emphasis on translating findings into a variety of useful formats for different stakeholders, including consumers.

The full report and this summary are available at www.effectivehealthcare.ahrq.gov/reports/final.cfm.



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Classification and Spectrum of Disorder

CP includes a spectrum of disorders of movement, posture, and coordination with heterogeneous etiologies. The diversity of the clinical features is reflected in multiple classification systems that include reference to type of motor dysfunction, body parts affected, severity, and functional abilities. Further classification is by severity level (mild, moderate, severe), and gross motor function, which reflects the functional capabilities of the affected.^{10,11} Developed in the late 1990s, the Gross Motor Function Classification System (GMFCS) outlines a standardized system for classifying motor function based on constructs of disability and functional limitation.¹² The GMFCS includes levels that reflect abilities ranging from walking without limitations (level I) to severe head and trunk control limitations requiring extensive use of assisted technology, physical assistance, and a wheelchair (level V). Table A summarizes criteria used in widely accepted classification systems.

The epidemiologic Oxford Feeding Study reported significant correlations between severity of motor impairment and feeding problems including choking, underweight, prolonged feeding times, vomiting, and need for gastrostomy feeding (p values typically <0.005).¹³ Although CP is a motor disorder, many children and adults with CP are affected by other developmental disabilities, including intellectual disability, impaired vision and hearing, language and behavioral disorders, and epilepsy.^{11,14,15} Survival and quality of life vary across the spectrum of CP, but both are associated with severity and functional disabilities, as well as comorbid conditions.¹⁵

Feeding Difficulties and Interventions

Individuals with CP frequently have feeding and swallowing problems that may lead to poor nutritional status, growth failure, chronic aspiration, esophagitis, and respiratory infections. Across the cerebral palsy spectrum, poor nutritional status is caused by distinct pathways ranging from inadequate intake, oral dysphagia, oral-pharyngeal dysphagia, gastroesophageal reflux, chronic aspiration, and behavioral etiologies. Some patients with oral-pharyngeal dysphagia and gastroesophageal reflux (GER), particularly those with severe CP, are also at risk for recurrent aspiration, which can lead to chronic pulmonary disease. Patients with feeding difficulties range from those with self-feeding skills to populations with severe disability (GMFCS V) who require extensive use of assisted technology and are dependent on others to feed them. Caregiver burden is a significant concern as the

feeding process may require considerable time and may be associated with stress and caregiver fatigue¹⁶; stress and fatigue may in turn affect the feeding process.¹⁷ A number of feeding and oral-motor intervention strategies have been developed to address difficulties with sucking, chewing, swallowing, and improve oral-motor skills. Strategies include oral sensorimotor management, positioning, oral appliances, food thickeners, specialized formulas, and neuromuscular stimulation. These interventions address different aspects of feeding difficulties, reflecting the range in specific problems associated with feeding and nutrition in CP. Sensorimotor techniques seek to strengthen oral-motor control and counteract abnormal tone and reflexes to improve oral feedings, and typically require months of daily application. Positioning techniques address poor postural alignment and control that exacerbates swallowing difficulties, and include stabilizing the neck and trunk. Positioning interventions are individualized and often guided by video-fluoroscopy to optimize swallowing. Oral appliances have been used to stabilize the jaw, improve sucking, tongue coordination, lip control, and chewing. Multiple approaches may be used in children with growth failure. For children with moderate to severe aspiration or malnutrition related to oral-pharyngeal dysphagia and GER, surgical interventions with gastrostomy (tube feeding directly into the stomach) or jejunostomy tubes (tube feeding into the middle portion of the small intestine, the jejunum) and antireflux procedures are often deemed necessary to improve nutritional status and reduce risk of chronic aspiration.^{16,18}

No uniform decision pathway exists for deciding when a child should move from oral feeding to enteral tube feedings, but there is general consensus.¹⁹ If oral calorie intake is insufficient to maintain growth, there is increased risk or occurrence of aspiration into the lungs, or the level of work necessary to maintain adequate caloric intake orally by the individual and the caregiver is excessive, then a medical provider may recommend enteral tube feedings (see Glossary). The method of tube feeding is based on the likely time span needed for tube supplementation, the availability of an experienced surgeon, and specific symptoms of the child. For example, a child may be considered too medically fragile for surgery, so a nasal tube may be used for a time, which may be advanced beyond the stomach into the jejunum to reduce gastroesophageal reflux, then later replaced with a surgically placed tube. A gastric fundoplication may be included to reduce GER, if needed in the judgment of the surgeon.

Table A. CP classification systems used and understood by qualified medical practitioners*

Severity Level	Topographical Distribution	Motor Function	Gross Motor Function Classification System
<ul style="list-style-type: none"> • Mild: Child can move without assistance; his or her daily activities are not limited. • Moderate: Child will need braces, medications, and adaptive technology to accomplish daily activities. • Severe: Child will require a wheelchair and will have significant challenges in accomplishing daily activities. 	<ul style="list-style-type: none"> • Monoplegia/monoparesis means only one limb is affected. It is believed this may be a form of hemiplegia/hemiparesis where one limb is significantly impaired. • Diplegia/diparesis usually indicates the legs are affected more than the arms; primarily affects the lower body. • Hemiplegia/hemiparesis indicates the arm and leg on one side of the body is affected. • Paraplegia/paraparesis means the lower half of the body, including both legs, are affected. • Triplesia/triparesis indicates three limbs are affected. This could be both arms and a leg, or both legs and an arm. Or, it could refer to one upper and one lower extremity and the face. • Double hemiplegia/double hemiparesis indicates all four limbs are involved, but one side of the body is more affected than the other. • Tetraplegia/tetraparesis indicates that all four limbs are involved, but three limbs are more affected than the fourth. • Quadriplegia/quadruparesis means that all four limbs are involved. • Pentaplegia/pentaparesis means all four limbs are involved, with neck and head paralysis often accompanied by eating and breathing complications. 	<ul style="list-style-type: none"> • Spastic: Implies increased muscle tone. Muscles continually contract, making limbs stiff, rigid, and resistant to flexing or relaxing. Reflexes can be exaggerated, while movements tend to be jerky and awkward. Arms and legs often affected. Tongue, mouth, and pharynx can be affected, as well, impairing speech, eating, breathing, and swallowing. Spastic CP is hypertonic and accounts for 70% to 80% of CP cases. The injury to the brain occurs in the pyramidal tract and is referred to as upper motor neuron damage. • Nonspastic: Decreased and/or fluctuating muscle tone. Multiple forms of nonspastic CP are each characterized by particular impairments; one main characteristic is involuntary movement, can be slow or fast, often repetitive, and sometimes rhythmic. Planned movements can exaggerate the effect (known as intention tremors). Stress can also worsen the involuntary movements, whereas sleeping often eliminates them. An injury in the brain outside the pyramidal tract causes nonspastic CP. Due to the location of the injury, mental impairment and seizures are less likely. Nonspastic CP is divided into two groups, ataxic and dyskinetic. Together they make up 20% of CP cases. Broken down, dyskinetic makes up 15% of all CP cases, and ataxic comprises 5%. 	<p>The GMFCS uses head control, movement transition, walking, and gross motor skills such as running, jumping, and navigating inclined or uneven surfaces to define a child's accomplishment level. The goal is to present an idea of how self-sufficient a child can be at home, at school, and at outdoor and indoor venues.</p> <ul style="list-style-type: none"> • GMFCS Level I: Walks without limitations. • GMFCS Level II: Walks with limitations. Limitations include walking long distances and balancing, but not as able as Level I to run or jump; may require use of mobility devices when first learning to walk, usually prior to age 4; and may rely on wheeled mobility equipment when outside of home for traveling long distances. • GMFCS Level III: Walks with adaptive equipment assistance. Requires hand-held mobility assistance to walk indoors, while utilizing wheeled mobility outdoors, in the community and at school; can sit on own or with limited external support; and has some independence in standing transfers. • GMFCS Level IV: Self-mobility with use of powered mobility assistance. Usually supported when sitting; self-mobility is limited; and likely to be transported in manual wheelchair or powered mobility. • GMFCS Level V: Severe head and trunk control limitations. Requires extensive use of assisted technology and physical assistance; and transported in a manual wheelchair; unless self-mobility can be achieved by learning to operate a powered wheelchair.

© Copyright 2011, Law Office of Kenneth A. Stern, PLLC. All Rights Reserved. For more information about types and forms of cerebral palsy please visit MyChild™ at cerebralpalsy.org/about-cerebral-palsy/types/.

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Clinical Uncertainties

The goal for management of CP is to improve the quality of life for both the child and family, through interventions that maximize independence in activities of daily living, mobility, and nutrition. Guidelines have been published by the American Academy of Neurology on the use of pharmacologic treatment of spasticity in children and adolescents with CP.²⁰ However, there is a limited evidence base for the majority of interventions in CP, including those that address nutrition and growth.²¹ Despite a range of potential feeding interventions for patients with CP, synthesis is lacking on the efficacy, safety, and applicability of these interventions. Limited information is available on the impact on health outcomes, including quality of life. Existing reviews are limited in scope, and clinicians and families will benefit from consolidation of data for making clinical decisions.

Goals of treatment and measures of effectiveness may differ by type of CP (spastic or nonspastic), location of motor involvement (e.g., diplegia, quadriplegia), functional status, including ability to walk or sit, and degree of head and trunk control. Comorbid conditions, particularly intellectual disability (related to ability to monitor and maintain appropriate nutrient intake) as well as concurrent medications that potentially have gastrointestinal side effects may influence treatment outcomes. Different feeding interventions may perform differently across the spectrum of CP. For example, oral-motor interventions may be highly effective in populations with oral dysphagia with malnutrition. However, these same interventions could have less value in less mobile populations that are experiencing pharyngeal dysphagia with aspiration. Gastrostomy feeding may reduce aspiration during swallowing, but does not address aspiration of oral secretions, and could exacerbate GER.²²⁻²⁴ Additional interventions, such as positioning and caloric supplementation may still be needed. To examine the overall effectiveness of interventions intended to improve feeding and nutrition outcomes in CP, adequate characterization of the patient populations is essential. Additionally, the need for management into later life has increased, and the optimal interventions for adults with feeding difficulties are unknown.^{8,25}

Potential harms associated with feeding interventions include surgical complications, new or worsening GER, risk of aspiration, and mortality. Gastrostomy has been associated with excess weight gain.²⁶ The impact of antireflux procedures in addition to gastrostomy is relatively unknown. Finally, there is a need to understand the potential impact of feeding interventions on families

and caregivers as substantial caregiver time and training may be required.

Objectives

The goal of this review is to examine the effects of available interventions for feeding and nutrition problems that have been evaluated in individuals with CP.

Population

We included studies whose populations included at least 80 percent of participants with CP. We did not require any specific diagnostic information or approach.

Interventions

Studies assessed interventions falling into the broad categories of nonsurgical interventions, including behavioral approaches (positioning, oral appliances, oral stimulation, sensorimotor facilitation, and caregiver training) and nutritional interventions (food thickeners, caloric supplementation with formulas, vitamin supplementation, and altering food consistency) and surgical interventions (gastrostomy tube [g-tube], percutaneous endoscopic gastrostomy [PEG], jejunostomy, and fundoplication). Studies may have used combinations of approaches (e.g., behavioral plus nutritional interventions).

Comparators

Comparators included other nonsurgical approaches or no intervention compared with behavioral interventions or nutritional interventions (Key Questions 1a, 2a), oral feeding or nutritional and behavioral interventions compared with tube feeding (Key Question 3a), oral feeding compared with g-tube with fundoplication (Key Question 3b), and jejunostomy tube compared with fundoplication (Key Question 3c).

Outcomes

Intermediate outcomes included changes in growth status, including height, weight, skinfold status, limb length, and energy expenditure; improvements in swallowing; and need for surgical or nutritional intervention. Patient-centered and health outcomes included mortality, hospitalizations, days of antibiotics for aspiration, quality of life, patient and family satisfaction and stress, feeding time, physical and mental health of caregiver, and reflux episodes. We also assessed the harms of interventions, defined by the Agency for Healthcare Research and Quality (AHRQ) Effective Health Care program as all possible adverse consequences of an intervention, including adverse events (Figure A).²⁷

Key Questions

We have synthesized evidence in the published literature to address these Key Questions:

Key Question 1a. When compared with other nonsurgical interventions or no intervention, how effective are behavioral interventions, including positioning, oral appliances, oral stimulation, sensorimotor facilitation, and caregiver training, for improving nutritional state/growth, health outcomes, health care/resource utilization, and quality of life in individuals with CP and feeding difficulties?

Key Question 1b. Is the effectiveness of behavioral interventions modified by age, race, severity, functional status (e.g., GMFCS level), or initial nutritional status?

Key Question 2a. When compared with other nonsurgical interventions (e.g., positioning, oral appliances or stimulation) or no intervention, how effective are nutritional interventions (food thickeners, caloric supplementation with formulas, vitamin supplementation, and altering food consistency [e.g., pureeing]) for improving nutritional state/growth, health outcomes, health care/resource utilization, and quality of life in individuals with CP and feeding difficulties?

Key Question 2b. Is the effectiveness of nutritional interventions modified by age, race, severity, functional status (e.g., GMFCS level), or initial nutritional status?

Key Question 3a. What is the comparative effectiveness of tube feeding when compared with oral feeding or with nutritional and behavioral interventions in individuals with CP who present with feeding difficulties, including malnourishment, failure to thrive, aspiration, and excessive caregiver burden?

Key Question 3b. Among individuals with CP and feeding difficulties *with significant reflux*, what is the effectiveness of g-tube placement with fundoplication versus oral feeding for reducing reflux and for improving nutritional state/growth, health outcomes, health care/resource utilization, and quality of life?

Key Question 3c. Among individuals *who develop reflux after gastrostomy*, what is the comparative effectiveness of j-tube versus fundoplication for reducing reflux in the short term and achieving improvements in nutritional state/growth, health outcomes, health care/resource utilization, and quality of life?

Key Question 3d. Is the effectiveness of tube feeding modified by tube placement, age, race, severity, functional

status (e.g., GMFCS level), initial nutritional status, or continuous versus bolus feeding?

Analytic Framework

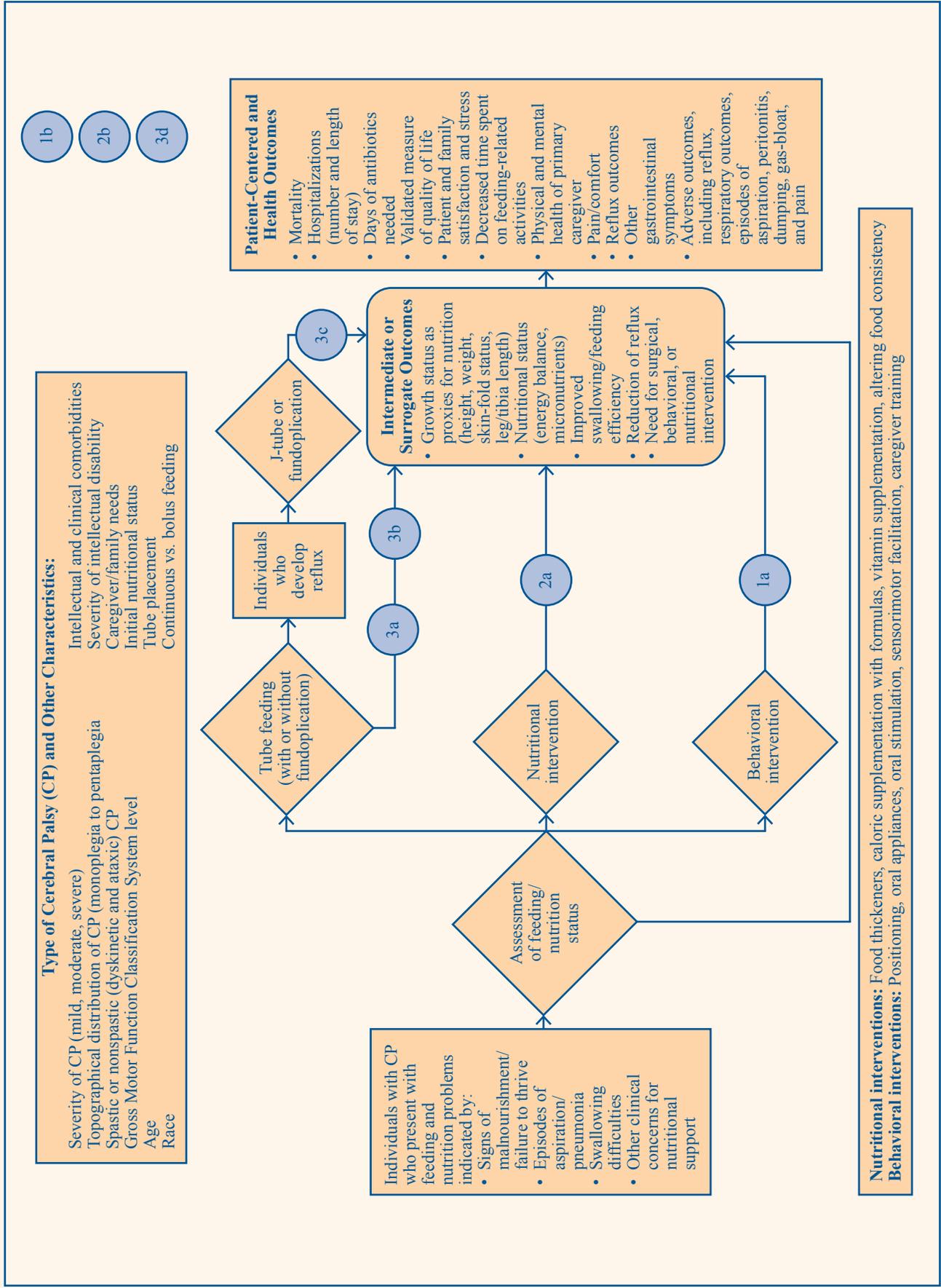
The analytic framework (Figure A) outlines the path of care for individuals with CP and feeding difficulties. There may be multiple indications among this population, including signs of malnourishment or failure to thrive, episodes of aspiration or pneumonia, swallowing difficulties, or other clinical concerns for nutritional support. Individuals typically undergo a feeding and nutrition assessment, which could be followed by a behavioral (Key Question 1a) or nutritional (Key Question 2a) feeding intervention or a combination of such approaches, or the placement of a tube for feeding (Key Questions 3a-c). Individuals with reflux may undergo tube placement with fundoplication to help alleviate reflux (Key Question 3b). Individuals without pre-existing reflux who undergo a tube placement may develop reflux following the procedure²²⁻²⁴ and require additional treatment via a jejunostomy tube or fundoplication (Key Question 3c). Possible intermediate or surrogate outcomes resulting from these interventions can include a change in growth status, improved swallowing, or various adverse effects. At this point on the pathway, individuals with CP may undergo another feeding and nutrition assessment followed by an alternative intervention. Patient-centered and health outcomes following the intermediate outcomes can include mortality, incidences of hospitalizations, antibiotic use, quality of life, patient and family satisfaction and stress, changes in time spent on feeding activities, physical and mental health of the primary caregiver, pain or comfort, and various adverse effects. Certain factors may influence the pathway at all stages and can include the type and severity of CP, age, race, intellectual and clinical comorbidities, severity of intellectual disability, and caregiver or family needs (Key Questions 1b, 2b, and 3d specifically address potential modifiers of treatment effectiveness). Numbers in circles within the diagram indicate the placement of Key Questions in relation to the treatment process.

Methods

Input From Stakeholders

The topic for this report was nominated in a public process. We drafted the initial Key Questions and analytic framework and refined them with input from key informants with expertise in child health and development, pediatric gastroenterology, occupational therapy, and

Figure A. Analytic framework



Note: Numbers in circles represent Key Questions.

neurodevelopment and developmental disabilities. After review from AHRQ, the questions and framework were posted to a public Web site. The public was invited to comment on these questions.

After reviewing the public commentary, we drafted final Key Questions and submitted them to AHRQ for review. During the topic development phase we identified a recent, rigorously conducted systematic review addressing behavioral feeding interventions.¹⁶ We thus structured the part of the review relevant to behavioral interventions as an update to the previous review. We convened a Technical Expert Panel (TEP) to provide input during the project on issues such as setting inclusion/exclusion criteria and assessing study quality. We identified key informants and TEP members through scanning recent research related to CP, reviewing stakeholders in an AHRQ-funded research exploration forum on CP,²⁸ and through discussions with our AHRQ Task Order Officer (TOO). All candidates were approved by the TOO after disclosure and review of potential conflicts of interest.

Data Sources and Selection

Data Sources

We searched key databases including studies related to surgical and nonsurgical interventions for promoting feeding and nutrition in individuals with CP: the MEDLINE[®] via the PubMed[®] interface, PsycINFO[®] (psychology and psychiatry database), the Cumulative Index of Nursing and Allied Health Literature (CINAHL[®]), OTSeeker, REHABDATA, and the Education Resources Information Clearinghouse (ERICsm). The appendixes of the full report include a description of the databases' content and breadth of coverage. Our search strategies used a combination of subject heading terms appropriate for each database and keywords relevant to CP and nutrition (e.g., cerebral palsy, enteral feeding). We also manually searched the reference lists of included studies and of recent systematic and narrative reviews and also invited TEP members to suggest potential citations.

Inclusion and Exclusion Criteria

We included all study designs except single case reports provided that studies reported on an intervention aimed at feeding/nutrition in individuals with CP. We excluded studies that:

- Were not original research
- Did not report information pertinent to the Key Questions
- Did not address treatment modalities aimed at outcomes of interest
- Did not include aggregate data (i.e., included only individual data for each participant) or data presented only in graphics/figures
- Were single case reports
- Were not published in English
- Were published before 1980.

Screening of Studies

Two reviewers separately evaluated each abstract. If one reviewer concluded that the article could be eligible, we retained it. Two reviewers independently read the full text of each included article to determine eligibility, with disagreements resolved via third-party adjudication.

Data Extraction and Quality Assessment

Data Extraction

A team member with methodologic expertise entered information into the evidence tables. After initial data extraction, a second team member edited entries for accuracy, completeness, and consistency.

Quality Assessment

Two reviewers independently assessed quality using quality assessment tools appropriate for the study design (Cochrane Risk of Bias tool for RCTs,²⁹ Newcastle-Ottawa scale for cohort studies,³⁰ a tool adapted from AHRQ Effective Health Care Program guidance for case series,³¹ and the AMSTAR tool for systematic reviews³²). The reliability and other characteristics of the Cochrane Risk of Bias tool, Newcastle scale, and AMSTAR have been previously assessed with positive ratings overall.³³⁻³⁸ We resolved differences through discussion, review of the publications, and consensus with the team. We rated studies as good, fair, or poor quality and retained poor studies as part of the evidence base discussed in this review. More information about our quality assessment methods is in the full report, and Table B describes the quality ratings.

Table B. Description of study quality levels

Quality Level	Description
Good	Good studies are considered to have the least bias and results are considered valid. A good study has a clear description of the population, setting, interventions, and comparison groups; uses a valid approach to allocate patients to treatments; has a low dropout rate; and uses appropriate means to prevent bias; measure outcomes; analyze and report results.
Fair	Fair studies are susceptible to some bias, but probably not sufficient to invalidate the results. A study may be missing information, making it difficult to assess limitations and potential problems. As the “fair quality” category is broad, studies with this rating vary in their strengths and weaknesses. The results of some fair-quality studies are possibly valid, while others are probably valid.
Poor	Poor studies are subject to significant bias that may invalidate the results. These studies have serious errors in design, analysis, or reporting; have large amounts of missing information; or have discrepancies in reporting. The results of a poor-quality study are at least as likely to reflect flaws in the study design as to indicate true differences between the compared interventions.

Data Synthesis and Analysis

Evidence Synthesis

Prior systematic reviews. When we identified published, high-quality systematic reviews addressing a Key Question that were largely up to date and relevant, we intended to cite and summarize these reviews as evidence and not extract data from the primary studies. One review met these criteria.¹⁶ We provide a summary of the methods of this review and overall findings in line with guidance in Using Existing Systematic Reviews to Replace de novo Processes in Conducting Comparative Effectiveness Reviews.³⁹

Primary research. For interventions not covered in existing systematic reviews, we extracted and synthesized data from primary studies meeting our criteria. The small number of the studies, the weak study designs and the heterogeneity in outcomes made a meta-analysis both inappropriate and unnecessary.

Strength of the Evidence

Prior systematic reviews. We used the included systematic review on behavioral interventions¹⁶ to assess strength of evidence for the literature included in the prior review, translating the assessment used in that review (see full report, Table 5) into levels used in the EPC program.

Primary research. We also assessed the strength of the body of literature for surgical studies included in the current review. The assessment of the literature is done by considering both the observed effectiveness of interventions and the confidence that we have in the

stability of those effects in the face of future research. The degree of confidence that the observed effect of an intervention is unlikely to change is presented as strength of evidence, and it can be regarded as insufficient, low, moderate, or high.

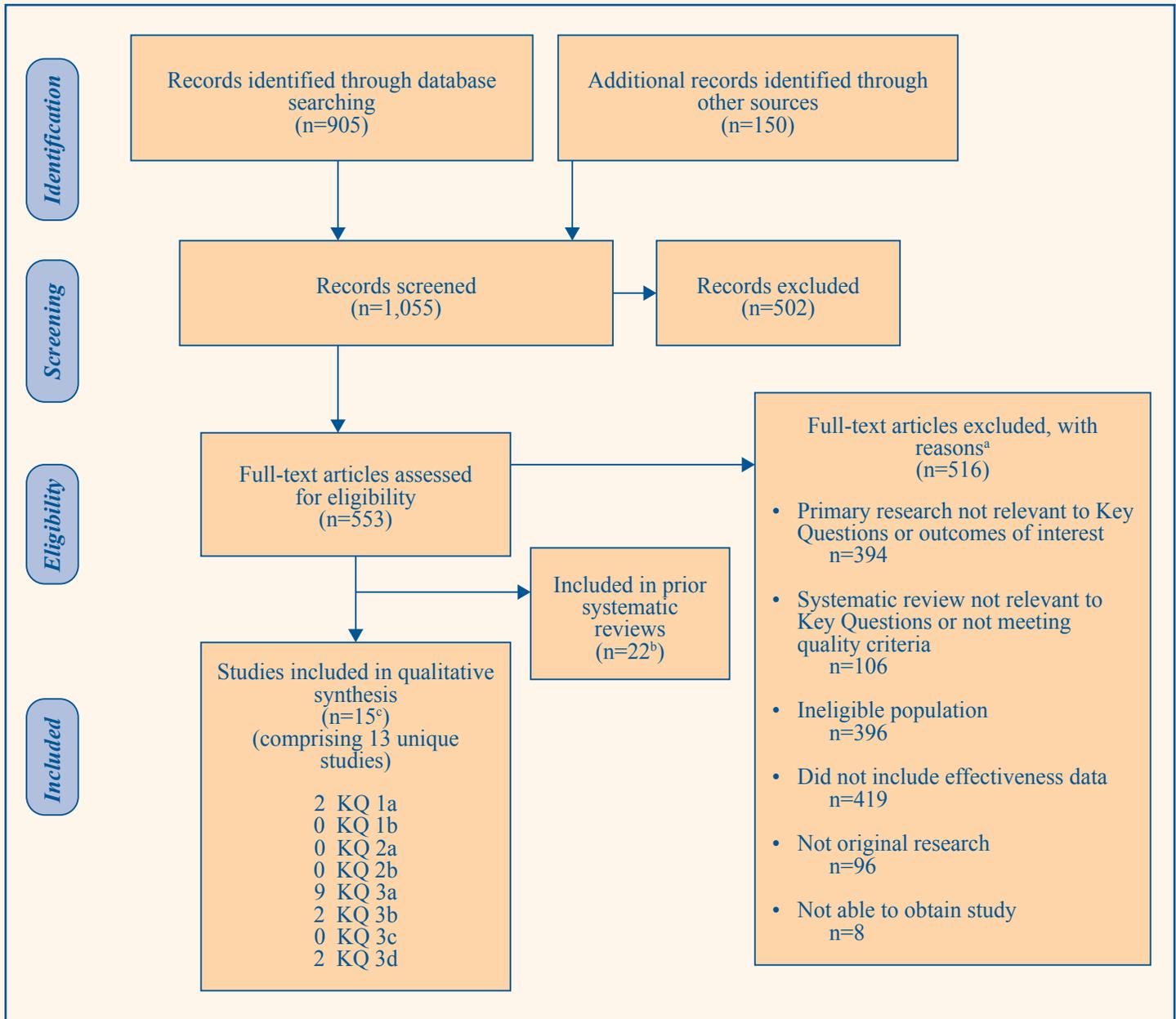
Methods for applying strength of evidence assessments are established in the AHRQ EHC Series Paper 5: Grading the Strength of a Body of Evidence When Comparing Medical Interventions⁴⁰ and are based on consideration of four domains: risk of bias, consistency in direction of the effect, directness in measuring intended outcomes, and precision of effect. Strength of evidence is assessed separately for major outcomes.

Results

Article Selection

Of the entire group of 1,055 citations, 553 articles required full-text review (Figure B). Of the 553 full-text articles reviewed, 15 articles (comprising 13 unique studies) met our inclusion criteria. This figure includes 12 unique primary research studies (reported in 14 publications) described in this comparative effectiveness review and one systematic review meeting our inclusion criteria. As indicated in Figure B, we were unable to obtain the full text of eight studies. Seven of these appeared from their abstracts to be narrative reviews, and one report, which may contain primary data, focuses on upper limb movement in CP. Thus, we do not feel that any of these papers would change our conclusions.

Figure B. Disposition of studies identified for this review



CER = comparative effectiveness review; KQ = Key Question; n = number

^aNumbers do not tally as studies could be excluded for multiple reasons.

^bThis number includes one study (Gisel 1994⁴¹) not explicitly referenced in the Snider review; the Snider review cites a later Gisel paper⁴² reporting on the same population.

^cThis figure includes 12 unique primary research studies (reported in 14 publications) described in the current CER and one systematic review meeting our screening criteria.

Summary of Findings by Key Question

Key Questions 1a-b. Effectiveness of Behavioral Interventions

Two studies assessing behavioral interventions met our criteria: one was a good quality systematic review.¹⁶ The primary literature updating the review consisted of one case series.⁴³ The systematic review, published in 2011 by Snider and colleagues, included 21 studies (5 RCTs), most with no more than 20 subjects, assessing interventions including sensorimotor approaches, positioning, oral appliances, altering food consistency, and feeding interventions, and largely addressing the outcomes of feeding efficiency and safety; overall, the review concluded that the evidence base was generally of poor quality that limited conclusions about effectiveness for all of the interventions.¹⁶ Effects of sensorimotor interventions were inconsistent, with one good quality study finding no effect, and smaller, less rigorous studies demonstrating improvements in efficiency and safety during feeding. A set of small case series of positioning found consistently positive effects, including reductions in food leakage and aspiration, but a larger, more rigorous study has yet to be conducted. One comparative study of the role of altering food consistency suggested that feeding could be made safer and more efficient by pureeing food. Oral appliances were associated with enhanced oral sensorimotor skills, but with only 2 RCTs, one of which was good, more research is needed. No effect of these appliances was seen on safety. Two small case series reported improvements in feeding efficiency, oral-motor behaviors and independence through the use of feeding devices. The small, short-term case series of a caregiver intervention⁴³ reported some improvements in oral-motor behaviors, caregiver stress, and number of chest infections, but it does not change the conclusions laid out in the prior review.

No studies were designed or powered to directly assess modifiers of the effectiveness of behavioral interventions (Key Question 1b). One study reported in the Snider review provides data separately for children with and without a history of aspiration.^{44,45} The study, reported in two publications rated as fair quality on the PEDro scale, reported improved eating efficiency and safety when eating pureed food for aspirating children after sensorimotor intervention, but no effect among those who did not aspirate.

Key Questions 2a-b. Effectiveness and Modifiers of Nutritional Interventions

No studies met criteria to address this question although pureed food was used in conjunction with positioning and sensorimotor interventions described in the review by Snider and colleagues.¹⁶

Key Question 3a. Effectiveness of Tube Feeding for Feeding Difficulties

The primary literature included six case series focused on assessing clinical outcomes after gastrostomy,⁴⁶⁻⁵³ one fair quality cohort study on the potential for overfeeding with gastrostomy that also included effectiveness data,⁵⁴ and one case series regarding the potential for gastrostomy to result in gastroesophageal reflux (GER).⁵⁵ Harms were also addressed in one case series assessing the effects of a low energy feed on the potential for overfeeding.²⁶

Evidence for the effectiveness of tube feeding (either g-tube or j-tube) comes from six case series and one prospective cohort study and one case series, designed to study the potential for overfeeding. All six case series assessing gastrostomy focused on severely impaired children and all reported significant increases in weight after gastrostomy, over six to 20 months. The most comprehensive case series⁵⁰⁻⁵² reported improvements on all weight and growth related outcomes (weight, head growth, linear growth, arm circumference and skinfold thickness), including closing the gap significantly with a normally developing reference population, and significantly more than would have been expected without intervention. Followup continued to 12 months postsurgery, with data available on 46 of the initial 57 children, and 6 unavailable due to loss to followup. The three other case series with growth data also reported significant pre-post increases in weight, but data on other measures were unreported or inconsistent. One case series also assessed health care utilization as a proxy for overall health and found the number of hospitalizations significantly reduced over the year following gastrostomy.⁵²

Two studies reported on QOL measures; in one,⁵¹ parental QOL improved significantly overall as rated on the SF-36 II, in tandem with decreases in feeding time. In the other, 98 percent of parents expected that their child's QOL (measured using a study-created tool based on questions from the validated CHQ-PF50 scale and a visual analog

scale) would improve with gastrostomy, but did not report that to be the case.⁴⁷

The studies included in this review were fairly short term and constrained by the requirement that they provide data both before and after surgery. Thus, harms of gastrostomy were limited to those in the fairly short term, and larger, retrospective studies that do report on harms data were excluded. Harms associated with tube feeding include surgical harms, infection, increased rates of reflux requiring further treatment, and potential overfeeding. Overall, rates of peritonitis were low, ranging from 2 to 5 percent, one study reported minor site infections at 59 percent and leakage at 30 percent. Deaths ranged from 7 to 29 percent, but were considered not to be related to gastrostomy in all studies. Mortality is high along individuals with CP, and it is impossible to know whether the observed deaths were causally related to treatment or to the course of the condition.

Three studies were specifically intended to analyze harms of tube feeding: one on the potential for tube feeding to induce reflux⁵⁵ and two others on potential to overfeed.^{26,54} Two studies found positive associations with the harmful outcome of interest (overfeeding or reflux).^{54,55} One study assessing the effects of a low energy feed, which increased weight without increasing fat mass, suggests that such formulas may have the potential to reduce the risk of overfeeding.²⁶ The clinical importance of these and other harms, relative to the potential danger of not intervening is unclear and likely must be assessed in the context of each individual patient and family. Clearly, surgical interventions can lead to increased weight gain; the degree to which harms outweigh those benefits likely depends on the starting point of the individual, family stressors, and the degree to which harms can be mitigated using appropriate feed and other approaches—an area that warrants continued research.

The frequent report of GER that develops after gastrostomy may be balanced by study investigators' observations that it is frequently managed medically. In terms of overfeeding, the two studies on this subject demonstrate that tube fed children may be at risk for obesity without careful attention to the content and quantity of their food products.

Key Question 3b. Effectiveness of Tube Placement With Fundoplication for Reducing Reflux

No studies directly compared the use of g-tube with fundoplication to oral feeding for the treatment of reflux.

One RCT including children with diplegic spastic or tetraplegic spastic CP compared two forms of plication (fundoplication versus vertical gastric plication),⁵⁷ and in one case series children with mixed and spastic forms of CP undergoing gastrostomy plus Nissen fundoplication showed improvements in reflux symptoms and weight gain but not episodes of pneumonia;⁵⁸ 30 percent of participants had recurrent reflux within 12 months of surgery. In the RCT, GER improved in both groups, with the Nissen fundoplication group having a significant decrease in the total number of reflux episodes, percentage of acidic pH, and longer reflux episodes, and the vertical gastric plication group showing a significant change in the pH measurement parameters.

Major complications were experienced by 14.3 percent of the children in each arm of the RCT, and minor harms included aspiration, urinary tract infection and pneumonia.⁵⁷ In the case series early complications (within one week of fundoplication) included pneumothorax, stomach ulcers, and cellulitis, all in patients undergoing laparoscopic fundoplication. Late complications (i.e., more than one week postfundoplication) included bloating, diarrhea (dumping syndrome), and intestinal obstruction; these events were all reported in individuals undergoing open fundoplication.

Key Question 3c. Effectiveness of J-tube Compared With Fundoplication

We did not identify any studies addressing this Key Question.

Key Question 3d. Modifiers of the Effectiveness of Surgical Interventions

We sought potential modifiers (age, race, severity, functional status, initial nutritional status, and continuous vs. bolus feeding) considered as important by our technical experts. Few studies addressed modifiers of effects of surgical interventions. Subanalyses were conducted in two case series^{46,48} to assess the degree to which age and type of procedure modified outcomes. In the first, children were divided into age bands of <2, 2 to 4, 5 to 7, 8 to 11 and 12 to 18. No age group included more than five children. Weight increased in all groups except ages 5 to 7, although this group had significant increases in triceps skinfold measurement. The very small size of each group, however, precludes any conclusion about age as a modifier.

The other retrospective case series of 57 individuals⁴⁸ reported that the highest proportion of individuals reaching weight for height were in the groups that had surgery

before age 2, had had their gastrostomy for at least 2 years or had fundoplication. One study assessing outcomes by the presence of fundoplication suggested that use of antibiotics and respiratory hospitalizations did not differ by whether the child had a fundoplication. The decision about whether or not to use fundoplication, was made clinically and not for research purposes.⁵² One series evaluating the g-tube with fundoplication found no difference in outcomes associated with laparoscopic versus open approach.⁵⁸ The intent of the study was not to compare the two approaches; rather, the clinical team changed their approach during the course of the study.

Discussion

Feeding and nutrition problems are common among children with CP and have significant health implications. Some patients with oral-pharyngeal dysphagia and GER, particularly those with severe CP, are also at risk for recurrent aspiration which can lead to chronic pulmonary disease. Patients with feeding difficulties range from those with self-feeding skills to populations with severe disability (GMFCS V) who require extensive use of assisted technology and are dependent on others to feed them. Indeed, chronic pulmonary disease related to aspiration is a leading cause of death among patients with severe CP.⁵⁹⁻⁶²

Ultimately, few data exist to guide care. Our analysis of the behavioral literature consists of a summary of a good quality systematic review published in 2011, updated with one new case series evaluating a caregiver training program that is not manualized (documented in a manual so that it can be replicated). The surgical literature consists of a total of 11 studies meeting our criteria; studies were largely case series. One prospective cohort study focused primarily on harms.

Across all interventions, the study populations are almost exclusively children with severe CP; when it is assessed populations generally meet criteria for level IV or V of the GMFCS. Although study populations are generally assessed on overall severity (e.g., GMFCS) and weight, the use of other measures for growth and nutrition, and explicit characterization of the feeding challenges in the study population is lacking. Surgical outcomes data are available for fewer than 300 children, and only one cohort study provides comparative data comparing surgical with oral interventions for any population of CP. Of note, those studies that do provide data on weight gain do so against reference populations of typically developing children. These are likely not appropriate reference standards; improvement in z-scores among children with CP may

very well be clinically meaningful even if these children do not approach weight standards for the reference group.

Key Findings and Strength of the Evidence

We used the included systematic review on behavioral interventions to assess strength of evidence, translating the assessment used in that review into levels used in the EPC program. Behavioral studies included in the prior review¹⁶ were small, typically short-term, and typically conducted using pre-post designs subject to bias. The authors of the systematic review used a modified Sackett approach to assess the strength of the body of evidence (see Table 5 in the full report). We have translated those assessments into EPC program equivalents in Table C.

Strength of evidence for behavioral interventions ranges from low to moderate. The moderate rating for the positive effects of oral appliances on sensorimotor outcomes is based on one good and one fair quality RCT and additional supporting studies of varying designs. Effects on eating efficiency and swallowing were not consistent, and the small sample sizes suggest imprecision. The low strength of evidence for positive effects of positioning, altering food consistency, and feeding devices on all outcomes is due to the lack of RCTs and generally small sample sizes. Studies typically reported some positive effects on mealtime length and eating efficiency; however, rigorously conducted studies are lacking. The strength of the evidence for the effects of oral sensorimotor interventions and oral appliance on feeding safety and efficiency is insufficient based on a paucity of rigorous studies.

Longer term studies are lacking across all interventions; thus, the durability of effects is not clear. Studies also did not consistently assess harms, though aspiration and swallowing difficulties, which may be related to the underlying condition as well as the intervention, are reported in some. Overall, more data on greater numbers of participants, including adults as well as children, are needed to understand the effectiveness of behavioral approaches.

We also assessed strength of evidence for six primary outcomes associated with feeding tubes in comparison with oral feeding, and for feeding tubes with fundoplication to address reflux: changes in growth outcomes; respiratory outcomes, including reflux; quality of life; long term morbidity and mortality; and harms (Tables D–E). We found the evidence to be insufficient to low for all outcomes. The low strength of evidence for the effects of gastrostomy on increasing growth measures, including weight, is based on a clearly significant effect measured in five case series and one prospective cohort

Table C. Strength of the evidence for behavioral interventions assessed in Snider review¹⁶

Intervention	Outcome(s)	Level of Evidence (Sackett)	EPC Equivalent Strength of Evidence (Direction of Effect)
Oral sensorimotor interventions	Increased feeding safety and efficiency	4 (conflicting)	Insufficient Inconsistent evidence and a paucity of comparative studies. Poor quality studies had positive results; whereas those with more rigor showed no effect, but may have been underpowered.
Positioning	Increased feeding safety and efficiency	2b (limited)	Low No RCTs, but positive results consistently observed in other study designs. Studies were small, and therefore imprecise.
Altering food consistency	Increased feeding safety and efficiency	2b (limited)	Low One experimental study that was of adequate size showed some positive effects on increasing feeding safety and efficiency.
Oral appliances	Enhanced oral sensorimotor skills	1b (moderate)	Moderate One good RCT, one fair RCT, and additional supporting studies of varying designs. Better quality studies showed positive effects, but effects were not entirely consistent; small sample sizes suggest imprecision and rigorous studies should be replicated.
Oral appliances	Increased feeding safety and efficiency, generalized postural control	5 (no good evidence)	Insufficient Only studies of poor quality were available to assess feeding efficiency and generalized postural control.
Feeding devices	Increased feeding efficiency	2b (limited)	Low Consistently positive results in two non-RCTs of small sample sizes.
Feeding devices	Enhanced oral-motor behaviors	2b (limited)	Low Consistently positive results in two non-RCTs of small sample sizes.
Feeding devices	Increased independence	2b (limited)	Low Consistently positive results in two non-RCTs of small sample sizes.

EPC = Evidence-based Practice Center; RCT = randomized controlled trial

study and in a small number of children. Additional data are needed on greater numbers of children to better quantify expected effects, particularly in subgroups by severity and age, and to better understand the implications of observed harms. Long-term effects are unknown as data on mortality are short term only. Nonetheless, it is clear that, in children with significant feeding difficulties, most of whom present significantly underweight, tube feeding leads to weight gain. Evidence is currently insufficient to assess whether and to what degree fundoplication is effective specifically to treat children with CP who present with significant reflux.

Applicability of the Evidence

Applicability of Studies of Behavioral Interventions

Studies of behavioral interventions to date have been limited in scope and focus on a limited selection of outcomes of interest. Studies typically provided limited data on health outcomes including hospitalizations, antibiotic use, patient and family satisfaction and quality of life, measures of family stress, and pain/comfort. In addition to the recent systematic review from Snider and colleagues, we located one case series based in the home among child-caregiver pairs in Bangladesh.

Table D. Outcome, strength of evidence domains, and strength of evidence for feeding tubes (KQ3a)

Outcome	Study Type (Number Reporting Outcome)	Domains Pertaining to Strength of Evidence (SOE)				SOE (Direction of Effect)
		Risk of Bias	Consistency	Directness	Precision	
Growth measures (weight, height, skinfold) ^{26,46-50,53,54}	Case series (7) Prospective cohort (1)	High	Consistent	Direct	NR	Low (Increase in growth measures)
Respiratory outcomes ^{26,52}	Case series (2)	High	NA	Direct	NR	Insufficient
Parental quality of life ⁵¹	Case series (1)	High	NA	Direct	NR	Insufficient
Child quality of life ⁴⁷	Case series (1)	High	NA	Indirect	NR	Insufficient
Long term morbidity and mortality	None					Insufficient
Harms ^{26,46-48,51,54,55}	Case series (6) Prospective cohort (1)	High	Consistent	Direct	NR	Low (Increased potential for overfeeding and reflux)

KQ = Key Question; NA = not applicable; NR = not reported; SOE = strength of evidence

Table E. Outcome, strength of evidence domains, and strength of evidence for fundoplication (KQ3b)

Outcome	Study Type and Number Reporting Outcome	Domains Pertaining to Strength of Evidence (SOE)				SOE
		Risk of Bias	Consistency	Directness	Precision	
Growth measures (weight, height, skinfold) ⁵⁸	Case series (1)	High	NA	Direct	NR	Insufficient
Reflux outcomes ^{57,58}	RCT (1); Case series (1)	High	Inconsistent	Direct	NR	Insufficient
Quality of life	None					Insufficient
Long term morbidity and mortality	None					Insufficient
Harms ^{57,58}	RCT (1); Case series (1)	High	Consistent	Direct	NR	Insufficient

KQ = Key Question; NA = not applicable; NR = not reported; RCT = randomized controlled trial; SOE = strength of evidence

The study focused on caregiver training related to diet, food consistency, appropriate utensils, and postural and physical support for positioning and feeding. Evidence from this study is likely primarily applicable to younger children who are able to eat at least some foods orally. The approach studied may not closely match interventions available in practice as it was conducted in the home setting, which is likely highly variable, and was not well described. Thus, individuals wishing to infer the potential results of clinical practice based on the available research need to assess carefully the degree to which the study methods matched those available and used in practice. Ultimately, the effectiveness of behavioral interventions within and outside of this limited sample and setting is currently unknown.

Applicability of Studies of Surgical Interventions

All of the studies of surgical interventions focused, appropriately, on severely impaired individuals, generally GMFCS levels of IV or V. Those studies that provided data to characterize the participants indicated that children in the studies had experienced substantial lack of growth for up to 12 months prior to intervention. Participants were followed for 6 months to over a year, and studies assessed outcomes of interest to clinicians and caregivers of individuals with CP, including changes in measures of growth, hospitalizations, and chest infections. The two studies of fundoplication for reflux similarly included children, but their level of functional impairment was not clearly described. Studies were not designed to assess subsets of individuals as defined by types of feeding disorders or specific surgical intervention.

Future Research

The study of feeding and nutritional interventions for individuals with cerebral palsy is a nascent field, but certainly one that is growing. Rigorous, comparative studies of behavioral and nutritional interventions need to be conducted; good RCTs are largely missing from the literature. Nonetheless, current research is available to provide potential directions for study. For example, studies of sensorimotor interventions currently provide conflicting evidence and more rigorous evidence is needed to answer the open question of whether they can be effective at improving outcomes. Studies of positioning are also warranted. Studies should also compare behavioral interventions with one another, with extensive characterization of the participants to better understand what works for which patients. Foundational research is needed to establish the most appropriate, patient-centered outcomes that are important to families of individuals with

CP. The degree to which improved changes are considered target outcomes by families is not well established. It is also not clear whether short-term outcomes translate to longer term health outcomes. We note that there is a complete lack of studies designed or powered to identify modifiers of effectiveness of the behavioral interventions.

The ethics of conducting comparative surgical studies or studies of nutritional interventions in the absence of appropriate comparison groups may preclude rigorous comparative designs. Case series can be conducted in ways that move them closer to providing effectiveness data; in addition, well-developed registries may provide a source of data for observational study designs. Of particular importance is the need to conduct large enough studies to fully characterize both participants and interventions so that the question of whether treatment approaches are better for individuals who, for example, aspirate or do not aspirate, can be answered. Patients with cerebral palsy are heterogeneous in many ways, including severity and comorbid conditions; rigorous subgroup analyses are needed to obtain data for targeting treatment. Furthermore, they and their families already experience substantial burden in terms of health care and other stressors. Recruitment and retention is likely to be a challenge, and may be a reason for the relatively poor evidence base to date.

In both types of interventions, data are absent on the role of feeding interventions for adults with CP. In addition to the interventions included in this review, it is necessary to consider the nutritional makeup (energy composition) of the food products themselves. Prospective, comparative studies should be carefully conducted to determine what type of nutrition is appropriate for obtaining positive health outcomes without inducing excessive weight gain.

Considerable uncertainty remains concerning harms over both the short and long term. Harms associated with feeding interventions have not been thoroughly reviewed in prior systematic reviews, and observational studies continue to raise questions about the risks and benefits of surgical interventions for children with severe CP and feeding difficulties.

Implications for Clinical and Policy Decisionmaking

The effectiveness of feeding and nutrition interventions for individuals with cerebral palsy remains largely unknown, with strength of evidence not exceeding moderate for any intervention. Nonetheless, clinical decisionmakers can use this review to understand what interventions are available,

what outcomes have been seen, and, to some degree, to balance potential harms. When a child has a severe feeding disorder, is unable to consume adequate nutrition, and is affected by frequent aspiration and pneumonias, the health outcomes can be dire. Understandably, treatment decisions must be made, even with inadequate evidence. Parents and providers contemplating gastrostomy can use the review to help understand potential effects on their quality of life and that of the child, potential harms that may occur, and potential tradeoffs related to social functioning. They should do so in light of the severity and other issues facing the individual child and family. Of note, nonclinical considerations may include family stress and pressures related to providing optimal care for the individual child. Stressors associated with caring for a severely disabled child and the potential impact of feeding interventions on the relationship with the child should not be underestimated and may play into decisionmaking along with the limited clinical evidence available. Ideally, this review will help policymakers and researchers understand what types of studies are essential to lead to more informed clinical decisionmaking.

Conclusions

Evidence for behavioral interventions for feeding disorders in cerebral palsy ranges from insufficient to moderate. Some studies suggest that sensorimotor interventions such as oral appliances (moderate strength of evidence) and positioning (low strength of evidence) may be beneficial, but there is a clear need for rigorous, comparative studies. Evidence for surgical interventions is insufficient to low. All studies to date demonstrate significant weight gain with gastrostomy. Results for other growth measures are mixed, and substantial numbers of children remained underweight, although given a lack of appropriate reference standards for the CP population, these results should be interpreted cautiously. Considerable uncertainty remains concerning harms over both the short and long term. Harms with gastrostomy can be common, and include overfeeding, site infection, stomach ulcer, and reflux. Mortality rates range from 7 to 29 percent. Longer-term, comprehensive case series are needed to understand potential harms in the context of benefits and potential risk of not treating.

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Glossary

Bolus feeding. Method of delivering enteral feedings using a limited amount of nutritional product administered through a tube into the stomach over a span of 15–30 minutes several times per day; not usually recommended for persons with a jejunostomy tube as the intestine cannot hold the same volume that the stomach can.¹

Castillo-Morales Device. Oral device including removable plates positioned on the upper jaw and including stimulatory elements to promote normal tongue and lip movements.

Continuous feeding. Method of delivering nourishment that involves the drip of formula by gravity or assisted by a pump in an ongoing manner over a specified number of hours into a gastrostomy, jejunostomy, or gastrojejunal tube.¹

Endoscopy. Procedure in which an instrument containing a camera is inserted into the gastrointestinal tract to visualize organs. This procedure is one of the methods used in the percutaneous placement of gastrostomy, jejunostomy, or gastrojejunal tubes.¹

Enteral feeding tube. Feeding device placed into the stomach or jejunum (middle section of the small intestine) through which formula, fluids, and/or medication are given to a person as an alternative to oral feeding.¹

Fundoplication/Nissen Fundoplication. Surgical procedure performed for the management of GERD. During the Nissen fundoplication, the upper part of the stomach is wrapped around the lower esophageal sphincter (the ring of muscle at the bottom of the esophagus that acts like a valve between the esophagus and stomach) to strengthen the sphincter and prevent acid reflux. The Nissen fundoplication may be performed using a laparoscope, an instrument that is inserted through tiny incisions in the abdomen, and uses small instruments to hold a camera to look at the abdomen and pelvis, which is less invasive and promotes faster recovery but requires more technical skill.^{2,3}

Gastroesophageal reflux (GER). Occurs when stomach contents reflux, or back up, into the esophagus (tube that connects the mouth to the stomach) during or after a meal. GER occurs when the lower esophageal sphincter opens spontaneously, for varying periods of time, or does not close properly and stomach contents rise up into the esophagus. GER is also called acid reflux or acid regurgitation, because digestive juices—called acids—rise up with the food. When refluxed stomach acid touches the lining of the esophagus it may cause a burning sensation in the chest or throat (heartburn or acid indigestion).²

Gastroesophageal reflux disease (GERD). More serious form of gastroesophageal reflux (GER); when acid reflux occurs, food or fluid may rise into the back of the mouth and could then fall down into the lungs, causing respiratory symptoms. Some degree of GER is considered normal, but persistent reflux that occurs more than twice a week or causes symptoms is considered GERD, and it can eventually lead to more serious health problems.²

Gastrojejunal (G/J-tube). Type of tube for nutritional support that is inserted into the jejunum (the middle section of the small intestine) through an established gastrostomy. It is also referred to as a G/J-tube or transgastric tube.¹ This uses a double lumen tube with 2 ports or openings. The G tube opening empties into the stomach and can be used for medication and the

J-(jejunum) tube opening which empties into the small intestine can be used for feedings and water.⁴

Gastrostomy. Surgical procedure that creates an artificial opening in the stomach for the insertion of a feeding tube.⁵

Gastrostomy tube (G-tube) insertion. Placement of a feeding tube through the skin and the stomach wall, directly into the stomach (also called a G-tube). This tube helps with feeding and releases air from the stomach.⁶

Innsbruck Sensorimotor Activator and Regulator (ISMAR). Oral appliance designed to provide stability for the jaw to develop lip closure and tongue mobility, improving eating and drinking skills.⁷

Jejunostomy (J-tube). Surgically placing a feeding tube through the abdominal wall directly into a part of the small intestine called the jejunum. The feeding tube bypasses the stomach and delivers a special liquid food with nutrients directly into the jejunum.²

Nasogastric tube (NG-tube). Tube is inserted through the nose or mouth, down the esophagus, and into the stomach.⁶ Typically used for short term.⁴

Percutaneous endoscopic gastrostomy (PEG) tube insertion. Gastrostomy tubes can be placed under endoscopic guidance, using a much smaller incision (percutaneous endoscopic gastrostomy tube placement, or PEG). An endoscope is passed into the mouth, down the esophagus, and into the stomach. The surgeon can then see the stomach wall through which the PEG tube will pass. Under direct visualization with the endoscope, a PEG tube passes through the skin of the abdomen, through a very small incision, and into the stomach. A balloon is then blown up on the end of the tube, holding in place. PEG gastrostomy tubes avoid the need for general anesthesia and a large incision.⁶

Percutaneous endoscopic jejunostomy. A type of J-tube placement for nutritional support that occurs with the aid of endoscopy to visualize the jejunum so that a tube can be threaded through a small opening made in the abdominal wall into the jejunum. It is also known as a PEJ tube.¹

Glossary References

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Full Report

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