Physical Disabilities: Education and Related Services, 2014, 33(2), 53-70.
doi: 10.14434/pders.v33i2.13142

PDERS

ISSN: 2372-451X
http://scholarworks.iu.edu/journals/index.php/pders/index

Article

EDUCATIONAL IMPLICATIONS OF COSTELLO SYNDROME: THE TEAM APPROACH

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Abstract: Since the 1990s the number of children with disabilities placed within the general education classroom has steadily increased. Many of these children are provided special education services under the generic disability title "intellectual disability." Over the past decade, there has been a significant amount of research concerning rare genetic syndromes that result in separating developmental disabilities into categories using similar origins and characteristics. Costello syndrome is one of these rare syndromes. Although information is available to medical practitioners, it has seldom been communicated to educators. The authors have taken the available data and, using a team approach, made recommendations for involving medical and health professionals in educating students with Costello syndrome.

Keywords: *Costello syndrome; rare syndrome; multiple disabilities; educational implications*

Introduction

Over the past 30 years there has been a surge in research that resulted in the identification of rare syndromes within the category of intellectual disability (e.g., Williams, Angelman, Costello). With the identification of individuals with common characteristics and issues comes the possibility of developing a greater understanding of their shared needs and the involvement of professionals providing services that could improve their quality of life. Costello syndrome (CS) is one of the rare syndromes where there appears to be a lack of transfer of medically-focused information into the educational arena. The lack of current medical information becomes a significant issue as children with complicated disabilities enter inclusive general education classrooms. With the passing of the legislation and regulations such as Individuals with Disabilities Education Improvement Act (IDEA) of 2004 (P.L. 108-446) and No Child Left Behind Act (NCLB) of 2001 (P.L. 107-110), the general education environment has been identified as the primary placement for students with disabilities. This paper will introduce educators to the needs of children with CS and the team approach used to enhance their quality of life.

History of Costello Syndrome

In 1971, Dr. Jack Costello, a pediatrician in New Zealand, identified two non-related individuals as having a cluster of characteristics that might be a new syndrome. After publishing his findings in 1977, no further research was published on this possible syndrome until Der Kaloustian, Moroz, McIntosh, Watters, and Blainchan (1991) reported another individual with similar characteristics. This new syndrome was named Costello syndrome and was defined as "a distinct multiple congenital malformation syndrome characterized by postnatal growth retardation, distinctive face, lax skin, and developmental delay" (International Costello Syndrome Support Group, 2004). Not all children with CS evidence all the aforementioned characteristics, but all who receive this diagnosis typically have a constellation of these characteristics present for that identification to be made. For example, characteristics that may be most recognizable are deeply creased palms and feet, curly hair, loose skin known as cutis laxa, and facial characteristics of a wide mouth, thick lips, high forehead, and large earlobes (Macnair, 2004; Nasca, Strano, Musumeci & Micali, 2003; National Organization of Rare Disorders [NORD], 2002).

The decade of the 1990s was a time when cases of children with what is now considered CS were reported internationally and information on the syndrome expanded rapidly (Borochowitz, Pavone, Mazor, Rizzo, & Dar, 1992; Davies & Hughes, 1994; Der Kaloustian, 1993; Di Rocco et al., 1993; Fryns, Vogels, Haegeman, Eggermont, & Van Den Berghe, 1994; Izumikawa, Naritomi, Tohma, Shiroma, & Hirayama, 1993; Kondo, Tamanaha, & Ashimine, 1993; Philip & Mancini, 1993; Say, Gucsavas, Morgan,& York, 1993; Siwik, Zahka, Wiesner, & Limwongse, 1998; Teebi & Shaabani, 1993; Umans, Decock, & Fryns, 1995; van Eeghen, van Gelderen, & Hennekam,1999). During the 1990s, the name Costello syndrome became the accepted nomenclature for the disorder (Costello, 1996; Johnson et al., 1998; Martin & Jones, 1991; Zampino et al., 1993).

The number of identified cases of individuals with CS throughout the world is 200-300 with a reported prevalence ranging from 1 in 300,000 to 1 in 1.25 million (U.S. National Library of Medicine, 2014). The search for the cause of CS has continued over the intervening decades with the primary focus being genetic factors (Delrue, Chateil, Arveiler, & Lancombe, 2003; Estep, Tidyman, Teitell, Cotter, & Rauen, 2006; Johnson et al., 1998; Kim et al., 2007; Kutsche, 2003; Macnair, 2004; Madhukara & Kumaran, 2007; Maroti et al., 2002; Tartaglia, Cottere, Zampino, Geld, & Rauen, 2003). Presently the causation is seen as mutations in the HRAS gene. The HRAS gene provides instructions related to cell growth. In this case, "the overactive protein directs cells to grow and divide constantly" (U.S. National Library of Medicine, 2014) often resulting in cancerous tumors (Gripp, Hopkins, Doyle, & Dobyns, 2010; Gripp & Lin, 2012). According to McCormick et al. (2013), p.G12A or p.G12C HRAS genes have more severe characteristics than those with other HRAS mutations. Unfortunately, the dissemination of this expanding research base to the educational community has not occurred. For educators to plan appropriate programs for children with CS, medically related information and its implications for appropriate educational programming is needed.

Characteristics and Their Impact

For the individual with CS, it is necessary to recognize the diversity of his/her needs resulting from this syndrome. Because a person with CS may exhibit variations in type and severity of characteristics, each individual must be viewed as unique. Following is a discussion of frequently occurring characteristics and their implications for family members and school personnel who are involved in educational planning. It should be noted that there exists a continuum of severity within the syndrome and that not all characteristics are present in every person with CS. Thus, these variations can be addressed in the individualized educational plan (IEP) so as to specifically meet the needs of the *individual* with CS.

Physical, Medical, and Health Characteristics

The unique physical characteristics associated with CS may be the first indication that the infant has this rare syndrome. A number of external physical characteristics are frequently noted in children with CS and are present at birth, such as macrocephaly, prominent forehead and wide mouth, ulnar deviation, cutis laxa (loose skin) or redundant skin folds on the neck, arms, and legs. Many children with CS have vision issues identifiable at birth or later including strabismus (crossed eyes), nystagmus (moving eyes), or acute nearsightedness (Johnson et al., 1998; Proud, 2003). Joint laxity may be noted early in life and can, along with the child's ulnar deviation, cause or result in significant gross and fine motor concerns (Kawame et al., 2003; Lin, Harding, & Silberbach, 2004; Philip, 2002). This joint laxity may result in delays in walking with significant limitations in ambulation (King, 2003; Yassir, Grottkau, & Goldberg, 2003). In addition, some children with CS have hypotonia, or muscle laxness, (Umans, Decock, & Fryns, 1995) and may need splints to support joints and musculature. Shortness of stature is a common characteristic as well as the development of scoliosis or kyphosis as the child matures (Lin, 2003; Yassir, Grottkau, & Goldberg, 2003). Among other possible physical markers are Chiari malformation and the presence of rare tumors and/or cancers such as Rhabdomyosarcoma or bladder cancer (Delrue Chateil, Arveiler, & Lacombe, 2003; Gripp et al., 2002). Chiari malformations are structural defects in the bony structure that holds the cerebellum and part of

the brain stem. As a result, these parts of the brain are pushed downward into the funnel-like opening of the spinal cord (known as the foramen magnum) resulting in possible issues with the functions controlled by these areas of the brain and a blockage of cerebrospinal fluid (National Institute of Neurological Disorders [NINDS], 2014). Depending on the severity of the malformation, surgery to correct the spinal issue or insertion of a shunt system may be required.

The medical characteristics alone are not specific to a diagnosis of CS and, thus, cannot be used as the sole criteria for identification. For example, one of the earliest medical characteristics often is the presence of congenital heart defects including mitral valve prolapse, atrial or ventricular septal defects, and pulmonary stenosis (Hou, 2000; Lin et al., 2002; Waldburg, Buehling, Evert, Burkhardt, & Welte, 2004; Williams, 2014). Children with CS have normal birth weight that declines rapidly after birth for no apparent reason (Kawame et al., 2003; NORD, 2002). According to Kawame et al. (2003), this physical decline, known as failure to thrive, is seen as a result of sucking and swallowing concerns that frequently occur. Another health concern is the presence of gastro-esophageal reflux (Macnair, 2004). In cases of severe feeding difficulties, a naso-gastric tube, or G-tube, may be necessary and, in some cases, tube feeding may continue until the age of two (Philip, 2002).

The occurrence of seizure activity is often present in persons with CS (Kawame et al., 2003) and may occur at different times in the individual's life. For example, it may be present at birth, may originate later in life, may continue throughout life, may stop and never return or sometimes may return later in life (Delrue et al., 2003; Kawame et al., 2003). Regardless of the time of onset, it is a significant medical concern and should be treated by medical specialists.

As a result of the complex and varied nature of the characteristics of individuals with CS, educators and other professionals should be aware of how medical issues and health concerns change throughout the lifespan of the individual. As the physical, medical, and health issues differ according to the age of the child, the following discussion is organized by age rather than by characteristic.

Birth to 5-years-old. During infancy and early childhood (birth to age 5 years), parents and professionals will work as a team and must focus on the development, implementation, and ongoing revision of an Individualized Family Service Plan (IFSP) as required by IDEA. The child's complicated medical and health needs may require a diverse team of professionals including cardiologists, neurologists, orthopedists, and ophthalmologists who may provide the requisite medical care for the child. At this age, medical specialists have an important role that includes the provision of medical and health information to the family and other professionals who are working with the family. Occupational therapists (OT), physical therapists (PT), nutritionists, and speech-language pathologists (SLP) are important partners in the development of appropriate programs for infants and young children with CS. The nutritionist will determine methods for meeting the nutritional needs of those children who have the characteristics of failure to thrive or inability to eat. In conjunction with the nutritionist, the SLP has a critical role working with the child and his/her family to address issues of sucking, chewing, and swallowing. When muscle laxity is present and impacts the health and development of the child, the family might need the expertise of a PT to assess and determine the proper physical supports necessary for feeding. The OT may be needed to address issues with the family related to the selection and

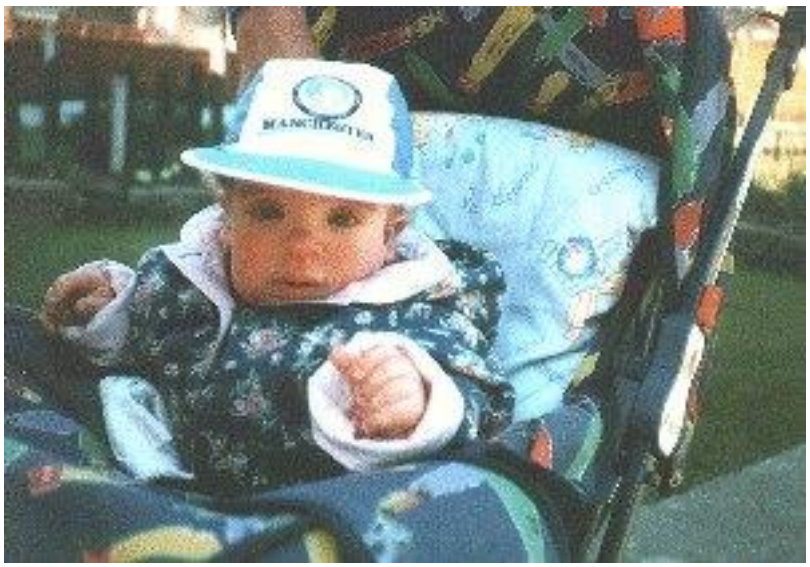

Figure 1. Helaina at 16 months

Figure 2. Helaina age 2-years

use of specialized equipment. In addition, OTs can assist the parents in identifying developmentally appropriate puzzles, blocks, peg boards, and wind-up toys for developing, strengthening, and refining the child's fine motor skills. The PT will also address any issues related to increasing the child's range of motion, balance, coordination, and locomotor skills including walking which is often delayed. If the child requires orthotics, splints, braces, or mobility aids, the OT and the PT can work with the orthopedist and the family in the appropriate selection and use of such items.

The early interventionist is an essential member of the team to work with family members to develop and/or increase the child's activities of daily living skills such as self-feeding and self-dressing and also assisting with the development of fine and gross motor skills, if they are delayed. To encourage fine and gross motor development the early interventionist, and possibly OTs or PTs, will work together to prepare a sequential motor development plan that can provide guidance for the family and others in identifying appropriate play activities for encouraging growth in motor skills and in developing a safe, but challenging environment for the child. If vision concerns are present and these concerns impact the child's ability to process sensory information visually, then the early interventionist may work with the family to create activities to assist in developing visual perceptual skills such as eye gaze, visual tracking, pattern perception, and depth perception.

Figure 3. Helaina at age 3-years

Age 6 to 12-years-old. Around the age of 6 and continuing until age 12, the child is considered an elementary-school aged child. As the child transitions into the inclusive classroom, orthopedic and vision issues may continue. As a result, orthopedists, neurologists, and ophthalmologists may continue their involvement with the transdisciplinary team. OTs and PTs will continue their involvement as needed, focusing on mobility within and around the school, appropriate playground access and activities, adaptation of physical education, and activities of daily living (ADL) when appropriate.

The SLP may continue providing services as needed in the areas of receptive and expressive language development and speech development. These services may include pull-out services for individual work on the formation of speech as well as language development. In addition the SLP may provide consultative services to the classroom teacher and to the parents in the form of information concerning therapeutic activities for implementation at home and within the classroom setting.

Medical issues continue for these children. Although neither general nor special education teachers act as providers of medical services, both have an essential role as monitors of the child's health. They also serve as reporters to the family and other medical professionals concerning health concerns that they may note in the classroom setting. Often classroom teachers are not experienced in recognizing the complex medical issues presented by children with CS but will need to become aware that some children will have possible physical activity restrictions per medical direction. Medical and health issues will vary significantly in children with CS, although common responses to these issues include physical restrictions.

As children with CS are often delayed in physical development, general and special education teachers need to address issues related to the child's visual perceptual skills, visual-motor skills, and fine and gross motor skills. Although there are many methods for meeting the needs of the individual child, adaptive physical education is one possible strategy. Children may also need modifications or adaptations in learning to print, to use scissors, to color, to run, and to climb steps. Minimizing the number or amount of copying and writing activities may be essential. For many children the use of circling or underlining answers in lieu of written answers may be advantageous. The use of computers with large font is helpful for those students who have developed the fine motor skills essential for using the keyboard.

If vision issues are present in conjunction with motor issues, the child's ability in the area of academics may be impacted; therefore, teachers will need to address these issues when planning school activities. When planning educational activities, the teacher should ensure that information is presented both orally and visually. Any directions or instructions that are written will need to also be presented with auditory input to reinforce the child's responses. When presenting written assignments such as worksheets, children with visual issues may need to have material that is not visually "crowded" with additional space between items.

Figure 4. Helaina 16th birthday

Age 13 to 21-years-old. As the child transitions into middle school then secondary school, medical and health issues continue. Teachers should be aware of the continuing complicated medical and health needs of adolescents with CS. Information needs to be shared concerning possible vision problems that may impact the student's ability to function in a classroom, possible orthopedic and medical issues that might result in his/her continued need for modifications in the physical environment, and potential curricular and instructional adaptations. Adaptations described for children ages 6 to 12 years can be useful for this age student as well. In addition, if there are physical or orthopedic concerns, then adaptive physical education might

Figure 5. Helaina as an adult

be an appropriate recommendation. Additionally, where mobility is an issue, a wheelchair may be needed if the student will need to travel any distance or need to move quickly.

Since the student is rapidly approaching the time of transition to adult services, both general and special education teachers will be involved in developing educational plans that address (a) the use of assistive technology, (b) the acquisition of appropriate leisure and recreational skills, (c) the development of ADL skills, and (d) pre-vocational skills. In selecting the skills to be taught, teachers should be sensitive to the adolescent's unique characteristics as well as future plans and community resources. To support the family members during this time of transition to postsecondary/adult services, teachers should provide information on community and state resources and the eligibility criteria for resources of specialized care.

Teachers should begin to talk with the parents and the student about individual medical and nutritional needs so that transition planning can occur. Particularly important is the need for the person with CS to become involved in his or her medical program to the greatest extent possible. The level of participation of the adolescent in his/her personal program is dependent on the ability of the adolescent. However, efforts toward self-regulation, self-care, and self-advocacy should be supported.

Cognitive Characteristics

Some of the cognitive characteristics of children with CS include developmental delay, intellectual disability, and memory deficits (Axelrad et al., 2004; Hennekam, 2003; Hou, 2000; Phillip, 2002; Proud, 2003). Receptive and expressive language differences related to cognitive abilities continue throughout the child's life (Johnson et al., 1998; King, 2003). In addition, the development of adaptive behavior and emotional and behavioral skills seem to be delayed (Axelrad, Glidden, Nicholson, & Gripp, 2004). However, a strength often noted in children with CS is their expansive, friendly personality (Hou, 2000), although it has been noted that irritability, hypersensitivity, and shyness might be present during the early years (Kawame et al., 2003).

As a result of the varied cognitive issues of individuals with CS, educators and other professionals must recognize these issues and how they may change throughout the lifespan of the individual. A discussion of cognitive issues and how educators may respond to them at different ages is discussed below.

Birth to 5-years-old. When addressing the cognitive characteristics of infants and young children with CS, important members of the transdisciplinary team, aside from the parents, are the SLPs and the early interventionists. As a result of tracheotomy tubes and naso-gastric tubes that are required by many infants, vocalizations and language development may be delayed. Both the SLP and the early interventionist will work with the family and the child to encourage the emergence of speech and language. As many children with CS appear to favor visual input, the use of sign language or other forms of communication such as the Picture Exchange Communication System or communication boards can assist in the development of the child's language (Harding, Lindsay, O'Brien, Dipper, & Wright, 2011).

Additionally, the early interventionist will work with the family to identify developmental landmarks and associated behaviors such as basic motor and perceptual skills and to provide activities to support and refine their development. As a result of the variability of developmental skills, intervention and support from other professionals (e.g., OT, PT, nutritionist) may be needed.

Age 6 to 12-years-old. At the beginning of elementary school, the child may still be functioning at the pre-academic level. SLPs will need to continue assisting with the child's speech development as well as with receptive and expressive language development. Teachers should remember that auditory and visual perceptual issues when paired with potential memory issues may impact early learning ability so classroom support is important.

Teachers and family members often state that the child with CS has an expansive personality or has "people pleasing behaviors." This expansiveness means that children often see everyone as a friend and may have difficulty controlling verbal outbursts such as calling out to friends during class. In addition, this may result in inappropriate physical behaviors such as hugging and kissing friends and teachers. These behaviors can become a safety concern as children may walk up to strangers or leave a safe environment without an understanding of the potential consequences of such behavior. All persons involved with the student should understand that these behaviors are

not necessarily under the child's control so he/she should not be punished, but encouraged to substitute more socially appropriate behaviors.

General and special education teachers will focus on educational planning that considers the child's cognitive level and its impact on academics as well as the child's language skills, memory skills, and motor skills. Learning to read has been identified as an area of significant difficulty for many children with intellectual disability, including those with CS (Allor, Champlin, Gifford, & Mathes, 2010; Allor, Mathes, Roberts, Cheatham, & Al Otaiba, 2014; Connor, Alberto, Compton, & O'Connor, 2014). A delay in the development of auditory perceptual skills may impact on the child's ability to learn to read through the phonics method. Alternative reading instruction such as a Rebus approach (Gately, 2006; Rani, 2012), a linguistic approach (Weaver, 2002), or a sight word approach (Browder, Wakeman, Spooner, Ahlgrim-Delzell, & Algozzine, 2006; Emenova & Behrmann, 2011) should be considered.

As a result of fine motor issues and possible vision issues, handwriting can become a major difficulty for children with CS. Handwriting instruction may need to be modified. Additionally, because of the child's difficulty with fine motor tasks, tasks requiring the use of scissors, pencils, instructional manipulatives, and art supplies may need to be modified or taught using adapted tools and materials. As children with CS have significant issues that impact their educational progress in school, the issue of system-wide alternative assessment must be addressed from the very beginning of their school program. For many of these children, portfolio assessment or other alternative forms of assessment should be considered in lieu of traditional forms.

Age 13 to 21-years-old. Beginning at age 13, plans focusing on transitioning students into secondary school and the community should be added to the educational program of adolescents and young adults with CS. Therefore, vocational rehabilitation counselors, transition specialists and other related service personnel may become a part of the transdisciplinary team. Families and schools might collaborate to assist the student in developing ADL skills for use at school and at home. Additionally, recreation and leisure activities might be taught at school and practiced during family time. When planning for the student's future as an adult, the issue of independent living and self-advocacy should be addressed with the parents and the person with CS. Training in self-advocacy and independent living skills will vary according to the abilities of the student. Teachers should target functional academics using appropriate academic modifications and compensatory equipment when needed. Pre-vocational and vocational skills become more important as the student nears graduation. All these areas must be addressed if the student with CS is to be prepared to function optimally in today's society.

Family Issues

For the family of a child with CS, every day brings a challenge. Without family support, a child with CS will not be able to function optimally in the home, school or community. However, the family also needs support. It is essential that families have information readily available when challenges occur. One potential source of support is the internet which can be used to connect families to current information through the government sites on Rare Syndromes (i.e., NORD or Orphanet). It can also provide a way for families to look at future potential challenges that their child with CS may encounter. In addition, the internet can be used to connect families of children

with CS through websites as well (e.g., International Costello Syndrome Support Group-UK and Costello Syndrome Family Network-United States). See Table 1 for a list of CS internet resources that are useful in locating information and connecting with families of individuals with CS. While this article is addressing only the needs of a child birth through 21 years, the individual with CS will continue to need support throughout the lifespan. When families come together, they can provide support to each other that can last a lifetime. Developing supports is a critical task for families and teachers and other professionals on the team should provide information, support, and assistance as needed.

Figure 6. Helaina with friends and family

Summary

Children and youth with Costello syndrome (CS) present a complex multi-systemic set of issues that must be addressed as the child moves through the educational system. Beginning at birth the child with CS and his/her parents will need support from a variety of individuals including medical professionals, early interventionists, occupational and physical therapists, and speech-language pathologists. As the child with CS moves through the educational system, the team members may vary but the complex systemic nature of this syndrome requires that support be provided for medical and educational issues throughout life. However, through this continuing team involvement, the individual with CS should have a greater opportunity to benefit from his/her school experience and successfully transition into the community. Likewise, families

benefit from developing supportive relationships with other families of individuals with CS which can last a lifetime.

Figure 7. Helaina age 20

Table 1

Websites Related to Costello Syndrome

Title of Site	Website Address
Archives of Pediatrics and Adolescent Medicine	http://archpedi.ama-assn.org/issues/v154n6/
Atlas of Genetics and Cytogenetics in Oncology and Haematology	http://atlasgeneticsoncology.org/Kprones/CostelloID10075.html
Costello Syndrome Family Network-United States	http://www.costellosyndromeusa.org
Genetics Education Center	http://www.kumc.edu/gec/support/costello.html
Genetics Home Reference	http://ghr.nlm.nih.gov/condition/costello-syndrome
International Costello Syndrome Support Group	http://www.costellokids.org.uk/welcome.htm
Medical News Today	http://www.medicalnewstoday.com/articles/221715.php

National Library of Medicine	**http://www.ncbi.nlm.nih.gov/entrez/query**
National Organization of Rare Disorders (NORD)	**http://www.rarediseases.org/**
Online Mendelian Inheritance in Man (OMIM)	**http://www.omim.org/entry/218040**
Orphanet	**http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Expert=3071**
Right Diagnosis	**http://www.rightdiagnosis.com/c/costello_syndrome/intro.htm**

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Author Note: A special thanks to International Costello Syndrome Support Group and to Colin Stone, its webmaster and founder and father of Helaina. The use of her photographs is important to our manuscript. Correspondence concerning this article should be addressed to Nikki L. Murdick, Department of Educational Studies, Saint Louis University, Saint Louis, MO 63103. Email: murdickn@slu.edu