



# **Educational Guidelines, IEPs, and School Services for Children with X & Y Chromosome Variations**

**Consensus-based recommendations from the  
AXYS Clinic & Research Consortium**

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## Introduction

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Students with X & Y chromosome variations (also known as sex chromosome aneuploidies, or SCAs) often require extra support from schools throughout the course of their education.

While SCAs affect approximately 1 in 500 live births,<sup>1</sup> they have long been significantly underdiagnosed due to limited physical symptoms. In fact, less than 10% of individuals with an SCA are currently diagnosed during childhood.

But recent advances in noninvasive prenatal testing are leading to more and more diagnoses before or soon after birth. This means we'll soon experience a dramatic rise in the number of students entering school with a diagnosis of an SCA. Educators must understand current research in SCAs and special education teams must be prepared with the best possible interventions and supports.

This document provides guidance on several issues related to educating students with SCAs. Our hope is that this resource will be useful for educators supporting students with SCA and for families navigating the education system.

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## Section 1 of 6: Genetics

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X & Y variations occur when a child is born with an atypical number of X or Y chromosomes, or both. Males typically have 1 X and 1 Y chromosome, while females are typically born with 2 X chromosomes. The most common X & Y variations are trisomy conditions, in which a male is born with an extra X or Y chromosome or a female is born with an extra X chromosome.

However, there are also instances when a child is born with 2 or 3 additional sex chromosomes ( such as XYY, XXY, or XXXX). When only some of the child's cells contain the extra X or Y chromosome, it is called mosaicism.

Prevalence rates vary significantly by condition, with an overall rate of 1 in 500 live births.<sup>1</sup> The following table lists the most common sex chromosome conditions.

Prevalence rates of X & Y chromosome variations in live births of males and females	
XXY (also called Klinefelter Syndrome or KS)	1 in 600 live male births
XYY	1 in 1,000 live male births
XXX (also called Trisomy X or Triple X)	1 in 1,000 live female births
XXYY	1 in 17,000 live male births

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## Section 2 of 6: Developmental considerations

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Students with SCAs are at increased risk of developing a number of educationally significant problems. The frequency and severity of problems differ by condition.

Children diagnosed prenatally tend to have better neurodevelopmental outcomes than those diagnosed later in life. This is because they are not diagnosed based on presentation with symptoms, but rather genetic testing and therefore represent the full spectrum of clinical severity in the SCA condition.<sup>5-7</sup> Children diagnosed postnatally represent the subgroup of those whose more significant neurodevelopmental challenges prompt the genetic testing that reveals their given SCA.

There is also significant variability in developmental outcomes within each diagnosed condition. This reflects the influence of other inherited genetic factors and environmental supports.<sup>8</sup>

### Medical and physical challenges

SCAs are not associated with a distinct physical birth defect as seen in other chromosomal trisomies (such as Down syndrome). However, a tall stature is common among all X & Y variations. Some students with SCAs will also present with widely spaced eyes (hypertelorism), skin folds covering the inner corner of eye (epicanthal fold), curved pinky fingers (clinodactyly), or low muscle tone.<sup>5,9</sup>

Boys with 1 or more extra X chromosomes (XXY, XXYY, XXXY) may have reduced levels of testosterone (called hypogonadism), small testes, and enlarged breasts (gynecomastia). However, early testosterone treatments may reduce these symptoms.<sup>10,11</sup> In adulthood, infertility is nearly universal for men with KS. But newly developed technology has allowed some men to become biological fathers through *in vitro* fertilization.<sup>12,13</sup>

### Developmental delays

More than half of all children born with an extra sex chromosome will show signs of developmental delay in early childhood.<sup>6,9</sup> About 60% will receive early intervention or early childhood special education (preschool) services to address or prevent delays.<sup>14</sup>

Early milestones may be late for infants and toddlers with SCAs, resulting in early speech, language, motor delays, or a combination of these. Reduced expressive language is the most commonly reported communication deficit, although receptive language is also compromised for many young children with SCAs. Low muscle tone and poor sensorimotor coordination can lead to lagging motor skills, including delayed walking.

## Learning challenges

General cognitive abilities for most students with SCAs are typically only slightly below average when compared to the general student population. However, verbal reasoning abilities are often significantly lower than nonverbal skills.<sup>5,15,16</sup> Those with XXYY and other variations in which there are more than 1 extra sex chromosome may present with more diminished cognitive ability, and approximately 30% are diagnosed with a mild intellectual disability.<sup>17</sup>

While most people with SCAs have cognitive abilities (IQ) within the average range, learning and academic challenges are common.<sup>18,19</sup> Current research indicates that a majority of students with SCAs will require some kind of school support plan (classroom accommodations or special education).<sup>20</sup> For school-aged children, reading accuracy, comprehension, and spelling are of particular concern. Prior studies indicate that up to 75% of children with SCAs will meet criteria for a diagnosis of language-based learning disability (such as dyslexia).<sup>15, 21-23</sup>

Several key cognitive processes have been identified across the lifespan as underlying skill deficits for individuals with SCA. These include weaknesses in:<sup>15</sup>

- Phonemic discrimination
- Verbal working memory
- Comprehension
- Fluency

In addition to language-based learning disabilities, research has also documented deficits in math problem solving and calculation for a subgroup of boys with KS and girls with Trisomy X.<sup>18</sup>

## Adaptive functioning

Despite having cognitive abilities within the average range, children with SCAs can have significantly more compromised adaptive skills than their non-SCA peers.<sup>24,25</sup> Educators

should be aware that these deficits are related to underlying challenges with language, executive function, and physiological differences and are not typically a sign of laziness or lack of motivation.

## **Attention and executive function**

Students with SCAs are at a considerably higher risk of developing attention-deficit/hyperactivity disorder (ADHD).<sup>25</sup> Prevalence rates of ADHD range from 36% for boys with XXY, 52% for girls with Trisomy X, 76% for boys with XYY, and 72% for boys with XXYY.

Most children with SCAs present with inattentive type ADHD, though boys with an extra Y chromosome can also display signs of hyperactivity and impulsivity. SCAs are associated with other executive function deficits in addition to attention concerns. Specifically, children with SCA have been shown to demonstrate deficits in the areas of inhibition, mental flexibility, and working memory.<sup>26</sup>

## **Mental health**

All SCA conditions are associated with an increased risk of mental health problems, especially anxiety and depression.<sup>26-28</sup> Females with Trisomy X have an increased risk of developing internalizing problems (such as anxiety, poor self-esteem, and social withdrawal) throughout their lives.<sup>5,29</sup> For boys with XXY, emotional issues related to social stigma, the negative impacts of having KS, and the worry of not being able to have children in the future may contribute to feelings of depression.<sup>30</sup>

## **Social skills**

Children with SCAs are at an increased risk for developing problems with social skills, including having a clinical diagnosis of autism spectrum disorder (ASD).<sup>5,24,31,32</sup> In particular, students with a Y chromosome aneuploidy are up to 20 times more likely to have ASD than their peers.

Supernumerary X chromosomes are also associated with an increased risk for autism, although symptoms may be less pronounced in these children. Further, students with SCAs who do not meet the cutoff criteria for an ASD diagnosis may still demonstrate immature social cognition and communication.<sup>33,34</sup>

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## Section 3 of 6: Available educational support in the U.S.

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### Special education

The Individuals with Disabilities Education Act (IDEA) is a federal law, which ensures that all students with disabilities ages birth to 21 have access to a free and appropriate public education (FAPE).<sup>35</sup> This education must take place in the least restrictive environment (LRE), meaning students should be educated alongside their typically developing peers as much as possible while still meeting their individualized goals. Goals and service delivery plans are developed through an individualized education program (IEP).

Students with SCAs do not automatically qualify for an IEP based on their genetic profile. However, many will require extra support in order to access grade-level curriculum.<sup>20</sup> Extensive research showing common learning challenges for students with SCAs (see above) suggests that special education teams should strongly consider evaluating for an educational disability if there are any learning concerns. A “wait and see” approach may prove harmful for these students, as their genetic condition can interfere with their ability to catch up with peers.

In order to receive special education support, a student must meet eligibility criteria for one of the following categories of educational disability.

**These are the education categories most frequently reported as “primary educational diagnosis” for children with SCA:**

- Autism
- Developmental delay/preschooler with a disability
- Emotional disturbance
- Intellectual disability
- Other health impairment
- Specific learning disability
- Speech or language impairment

### **Students can also meet eligibility criteria for one of these categories:**

- Deafness
- Deaf-blindness
- Hearing impairment
- Multiple disabilities
- Orthopedic impairment
- Traumatic brain injury
- Vision impairment (including blindness)

Qualification for an IEP is decided by a set of criteria determined by each state, or in some areas, individual school districts.

### **Individualized education program (IEP)**

The IEP is a legal document that guides an interdisciplinary team through the process of qualifying a student for special education and developing a comprehensive plan for specialized instruction.<sup>36</sup> At a minimum, the IEP team must include the child's parent, a general education teacher, and a representative from the school district.

#### **An IEP contains the following sections:**

- **Present levels of performance.** This section describes the child's current abilities, strengths, and weaknesses across all academic and functional domains. These may be discussed in an academic, social, or physical context, or a combination of these.

This section also explains how the student's disability affects their ability to access the general curriculum within the classroom. The team should base these descriptions on multiple sources when possible. Sources include teacher or therapist observations, the team's current goals for the child, and objective data (such as test results). The team can also include information from clinical psychoeducational or neuropsychological evaluations outside of school, but this isn't required.

This section should clearly describe the child's level of performance, with as much specific information as possible to help guide a more meaningful intervention plan. This includes information about the child's strengths and interests.



- **Individualized instruction and any related services.** This section outlines the special instruction a student will receive. It also outlines any related nonacademic services the student will receive (such as mental health support, speech therapy, occupational therapy, or physical therapy). These services allow the student to reach individualized goals and to participate fully in their education. Services included in an IEP are legally binding and schools are liable if they do not carry out the services as written.
- **Goals.** IEP goals are set annually and individualized to the student. Goals should be measurable, both functional and academic, and should emphasize enhancing the student's ability to access the general education curriculum.

Schools are not liable for a student's progress or lack of progress toward these goals. They are only liable for the specialized instruction and related services designed to meet these goals as outlined in the IEP.

- **Supports.** An IEP must also include descriptions of these 3 types of support the student will need to access the curriculum and demonstrate their understanding:
  - **Accommodations** (slight changes to the presentation of curriculum or student response options)
  - **Modifications** (changes in what content the student is expected to learn compared with grade-level curriculum)
  - **Assistive technology** (tools and devices to promote access and independence)
- **Least restrictive environment.** This section outlines any part of the student's day spent in a general education setting alongside typically developing peers.
- **Transition services.** Beginning at age 16, the IEP must include a plan for preparing the student for post-school goals. These include postsecondary education, job training, and independent living. This section also connects the student with agencies outside of the school system that the student will need for support in meeting these goals.

## IEP Checklist

Developing a solidly written IEP is critical for students with SCAs. Use this checklist to ensure your school team adheres to best practices throughout the IEP process.

- Parents are active partners in the IEP process. Parental input on a child's strengths and needs is critical.
- The IEP includes related services to support the child's communication needs, social interaction and adaptability, mental health, and motor and sensory needs. This will help ensure the child has the access skills needed for academic success.
- Goals are written in a way that allows the team to monitor each goal every day of the school year. This means that a report or update on progress would never read "we have not addressed this goal in class yet."
- Goals are specific and measurable. Each goal includes these 5 components:
  - Who (student)
  - Will do (the behavior/skill)
  - To what degree (criterion)
  - Under what condition (the supports)
  - In what length of time (time frame)
- The team has carefully considered the least restrictive environment for the student. This means the student will spend as much time as possible in a general education setting with the supports they need to succeed.
- If the student is 16 years old or older, the IEP includes an individualized transition plan detailing how the school will help prepare the student for life after high school. If the student is younger than 16, the IEP includes considerations for the student's long-term outcomes.

Each student with an SCA will need a transition plan based on their unique learning profiles, rather than their special education diagnosis. For instance,

many students with SCA will have cognitive skills in the average range but low adaptive skills. These students will need extensive support in order to achieve independent living, gainful employment, or postsecondary education or training. They may need a formal, structured transition program from age 18 to 21.

## 504 plans

Section 504 of the Rehabilitation Act (1973) is a federal civil rights law that requires agencies receiving federal funding to remove barriers to participation for individuals with disabilities.

Section 504 defines a student with a disability in a broader sense than IDEA. To qualify, students must have a physical or mental impairment that “substantially limits one or more life activities.” Students with SCAs may qualify for a 504 plan. This type of plan can provide accommodations, modifications, and some services. But unlike special education plans, 504 plans do not provide specialized instruction.

Examples of common 504 plan accommodations for testing and assignments for students with SCAs include:

- Extended time on assignments and exams
- Permission to use a keyboard for all assignments and assessments
- Scribing for exams and longer written classroom assignments
- Testing in a separate room
- Test instructions read aloud
- Assignment chunking
- Reduced homework requirements

Examples of additional common 504 plan accommodations for students with SCAs include:

- Passes for taking a break from the classroom as needed
- Preferential seating
- Speech-to-text and text-to-speech software
- Use of a private locker room/shower for P.E. class
- Check-ins with mental health staff
- An assigned notetaker in class

- Early access to material presented in class

## Multi-tiered systems of supports (MTSS), response to intervention, and problem-solving teams

Most school districts in the United States have moved toward a multi-tiered system of supports (MTSS).<sup>37</sup> This educational model emphasizes supporting all students with a goal of prevention. Because students with SCAs are at risk for developing educational disabilities, they will likely benefit from a well-developed MTSS process in the schools.

### The MTSS pyramid consists of 3 tiers of intervention:

- 1. Universal supports.** These are the evidence-based school and classroom-wide curricula that schools provide to all students. They include both academic instruction as well as universal social and emotional curricula (such as school-wide lessons about preventing bullying or positive behavioral expectations for all students).
- 2. Targeted supports.** These are added support measures that schools provide to students who need more help to meet grade-level standards. Targeted supports include efforts such as small groups focused on building specific reading skills, “lunch bunch” friendship groups, and “check-in check-out” behavior programs.
- 3. Individualized supports.** These are customized interventions that schools provide to students who require a higher level of support. These may be in the form of an IEP or a 504 plan. Individualized supports include efforts such as specialized instruction that specifically address a student’s unique cognitive profile.

Students should receive all levels of support that they need to meet grade-level standards.

### Examples of MTSS for 2 students with SCA

**Example 1: MTSS for a student with 47,XYX**

A student with 47, XYX receives daily support from a special educator for a reading disorder. The student's IEP clearly describes the needed supports.

The student's special education teacher has tailored a reading intervention to address his present levels of performance and move him toward his personalized goals for the end of the year. He also benefits from weekly targeted reading instruction with his classroom teacher in a small group. Further, he has access to a universal evidence-based school-wide reading curriculum each day.

**Example 2: MTSS for a student with Trisomy X**

A student with Trisomy X receives individualized accommodations for an anxiety disorder through a 504 plan. The student's IEP clearly describes the needed accommodations.

The student's accommodations include scheduled breaks and pressure passes that allow her to leave the class if she is feeling emotionally dysregulated. She also receives extended time on exams and reduced homework.

The student attends a tier 2 "lunch bunch" social skills group with the school counselor once a week for 10 weeks. Finally, she benefits from a school-wide positive behavior support program that encourages all students to demonstrate self-regulation and prosocial behaviors in the school.

**Response to Intervention (RtI)**

RtI is one method used to qualify students for special education with a specific learning disability (SLD) diagnosis within the MTSS framework.<sup>38</sup> It was developed as a problem-solving method to address long-standing concerns with the more traditional approach to SLD diagnosis (the discrepancy model), which some nicknamed the "wait to fail" model.

RtI is a data-based decision-making process in which a student receives a targeted intervention for specific skills and is closely monitored for skill development. Students who fail to "respond to interventions" at the level or rate required to close the gap with their same-aged peers may qualify for special education under the SLD category. The process has been expanded to include problem solving for social emotional and behavioral concerns as part of a larger MTSS process.

Students with SCAs who are struggling with academics or behavior may be referred by parents or a teacher to the school problem-solving team. Parents should be involved in the entire process to ensure the school team understands the student's risk factors related to the SCA diagnosis. Parents and teachers should bring data (such as work examples, test results, and attendance records) to the problem-solving meeting to help with making decisions.

**Example: A problem solving meeting for a student with an SCA**



**Calling the meeting**

Mr. Peterson referred his first grade student, Max, to the school's problem-solving team. He is concerned that Max is not making adequate growth in phonics compared with other students in his class. Max's parents have informed Mr. Peterson that he has a diagnosed SCA known as Klinefelter Syndrome (47, XXY or KS), but Mr. Peterson does not know much about this condition.

**Preparing for the meeting**

Mr. Peterson brings data to the meeting in order to guide the decision-making process. He brings along work samples from Max's phonics lessons and reading-level data from his weekly progress monitoring. He also brings along some comparison work from Max's classmates so the team can see the range of achievement in the class.

Max's parents bring several articles about Klinefelter Syndrome to help the education team understand this unique genetic condition. They also bring assessment data from when Max was in preschool and had received testing for early intervention. At that time, Max had shown some mild delays in his language such as speaking later than expected for his age.

**Advocating for the child's unique needs**

Max's parents explain to the team that Max's learning challenges are likely related to his diagnosis of Klinefelter Syndrome, and that research shows many students with this diagnosis will require some level of specialized support in school. They also advocate that the school intervene as early as possible, as Max may struggle to catch up if he falls much further behind his peers.

**Creating a personalized plan for intervention**

The problem-solving team uses the data brought by both the teacher and parents to create a plan for intervention. They document Max's current performance and set a reasonable goal for progress with a 6-week intervention. The goal is based on grade-level expectations and his current abilities.

The school's reading interventionist agrees to place Max in her daily phonics reading group for first graders who are struggling to learn letter sounds. The group meets for 20 minutes a day, 5 days a week. The interventionist will monitor Max's progress each week for 6 weeks.



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## Section 4 of 6: Educational services across the lifespan for students with SCAs

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### Early childhood (Ages birth to 5 years)

From birth to age 5, children receive early childhood services through 2 different management programs:

- **Early intervention (EI)** serves infants and toddlers ages birth to 3 years. Either Health and Human Services or the Department of Education manages EI, depending on the state.
- **Early childhood special education (ECSE)** serves children ages 3 to 5 years. The child's local school district manages ECSE services.

Parents of young children with a diagnosed SCA condition (including a prenatal or newborn diagnosis) can contact the local early intervention program to request an assessment or developmental screening. In some states, a diagnosis of SCA is sufficient to qualify a child for early intervention services. In others, the child must present with developmental delays in order to qualify.

The early intervention team will conduct a **multidisciplinary assessment** to screen for developmental delays. The assessment team should include multiple professionals with expertise in early development. This includes an early childhood special educator, a speech language pathologist, an occupational therapist, and/or an early childhood mental health professional (psychologist or social worker).

Due to the known risk factors for children with SCAs in the areas of language and motor development, the team should use specific standardized assessments to screen for delays regardless of any known deficits. It is critical for parents to identify developmental delays and begin therapies as soon as possible.

Research indicates that nearly 60% of young children with a diagnosed SCA condition receive some kind of EI therapy, and 39% receive more than one type of EI therapy. Children with tetrasomy or pentasomy conditions are significantly more likely to receive EI therapies (87%) than trisomy conditions (54%).

**U.S. families report their children with SCA receive the following EI therapies in order of frequency:**

- Speech therapy (44%)
- Physical therapy (33%)
- Occupational therapy (27%)
- Early developmental stimulation or early special education (22%)

For children under age 3 with delays, parents and the assessment team will work together to develop an **individualized family service plan (IFSP)**. This plan will include clear and measurable goals for the child, and will outline the plan for early intervention services based on the child's needs. This may include speech therapy, occupational therapy, physical therapy, or a combination of these. Therapies should take place in the home or another natural setting for the child, and parents will be equal partners in the intervention plan.

Starting at age 3, a child transitions from the IFSP managed by early intervention, to ECSE and an **individualized education program (IEP)**. More than half of preschoolers with SCAs receive ECSE services between the ages of 3 and 5.

Most students with SCA benefit from an **inclusive learning environment (also called a “least restrictive environment”)**, where they are educated alongside typically developing peers. In fact, 75% of young children with SCAs in the U.S. receive preschool services in an inclusive setting. In this environment, therapists employed by the schools provide therapies within the classroom to support the student's development in partnership with general and special education teachers.

**The most commonly reported preschool special education therapies received by children with SCAs are:**

- Speech therapy (45%)
- Early academic support (37%)
- Physical therapy (26%)
- Occupational therapy (23%)
- Social skills or behavioral supports (20%)

Each student's IEP provides individualized goals. Parents can and should communicate frequently with the school team to ensure the child is meeting their goals. Furthermore, parents should work with the school team to understand how the goals are being targeted and to learn about carryover activities they can do at home. Parent participation in developing the IEP at this age is critical.

## Elementary school (ages 5 to 10 years)

The K-12 school system is bound by the “**child find**” mandate of IDEA to seek out students with disabilities, evaluate them, and refer those who qualify for special education and support services. Parents can also request an evaluation for special education at any time. Teachers can refer a student for an initial special education evaluation, but parents must consent to the evaluation upon the referral.

Students with SCAs who enter kindergarten with a preschool IEP may continue special education services under the IDEA category of **developmental delay** or **preschooler with a disability** until their 9th birthday. Or, the team may choose to qualify the child under a more specific category such as **other health impaired (OHI)**, **speech language impairment (SLI)**, or **autism**.

For a student to qualify, the team must determine that a student needs special education services in order to receive a “reasonable benefit” from general education. At times, students with SCAs will not qualify based on their academic performance if their grades are still technically within the average range. Parents report hearing from teachers that their child is “not far enough behind” or their child is “at the low end of typical.” This can be a frustrating experience for families, as their child has likely had to work very hard through tutoring, behavior plans, and private therapies to keep their grades within the average range.

Executive function challenges and ADHD caused by SCAs may interfere with a child’s ability to keep up with peers efficiently or maintain energy in the classroom setting. Further, internalizing conditions, such as anxiety or social skills deficits, can cause students to appear shy or quiet in the classroom. Teachers may mistake such behavior for compliance, when in fact, the student is withdrawn and unable to concentrate and execute tasks without significant stress. The special education category OHI may fit well for such students whose limited “strength, vitality, or alertness” impedes their educational progress.

## Middle and high school (ages 11 to 18 years)

As in elementary school, middle and high schools are bound by **IDEA** in their searches to identify and provide services and supports for qualifying students. Students do not need to retest or requalify as they move into middle and then high school. Their established IEP or 504 plan will still be in place. The referral and testing processes remain the same throughout K-12.

Students with SCAs may find transitioning from the elementary setting to the middle school setting challenging. As students move through a typical K-12 system, the demands and expectations for organization, intrinsic motivation, and socialization increase. Problems with executive functioning, anxiety, and social deficits that the student compensated for in elementary school may become more difficult to manage. Students with SCAs can benefit from emotional-social therapy and organizational assistance in addition to other continuing accommodations from their younger years.

There are several things to consider for students approaching graduation and interested in college. School teams must clearly articulate expectations for graduation requirements, including standardized testing. Many school systems implement projects to be sure those students without passing scores on standardized tests still fulfill local standards for graduation. Additionally, accommodations can be provided for the PSAT and SAT, but applications need to be made by the student/family to the College Board.

## Transition to adulthood (ages 18 to 21)

Starting at age 16 (or around sophomore year in high school), the special education team should begin planning for the student's transition from the K-12 system. This will include an **individualized transition plan** in the student's IEP.

The unique cognitive profile of many students with SCAs (average IQ with lower adaptive skills and ability for independence) requires special consideration. Many traditional transition programs are reserved for young adults with intellectual disabilities and often leave out those with learning disabilities or more mild conditions.

Parents should actively encourage the special education team to begin the transition process early, creating a plan for next steps such as employment, vocational training, postsecondary education with educational supports, or a combination of these. Some students with SCAs may also need independent living skills instruction to achieve post-

school success. Transition plans can also connect students and families to community-based agencies that may be useful to the student after graduation. School social workers are often a resource for this information and can be a helpful partner at this stage in a student’s educational career.

<b>Educational services for students with SCAs</b>			
<b>Service</b>	<b>Description</b>	<b>Provided in response to</b>	<b>Appropriate age group</b>
<b>Speech language therapy</b>	Therapy to address communication delays or disorders that impact a student’s educational performance in school.	<ul style="list-style-type: none"> <li>● Receptive (following directions) language delays</li> <li>● Expressive (communicating wants/needs) language delays</li> <li>● Concerns about articulation</li> <li>● Social communication difficulties</li> <li>● Voice issues</li> <li>● Stuttering</li> <li>● Other speech and language problems</li> </ul>	All ages
<b>Occupational therapy</b>	Therapy to assist students in accomplishing the tasks of daily living, such as handwriting or paying attention in class.	<ul style="list-style-type: none"> <li>● Fine motor concerns</li> <li>● Sensory problems</li> <li>● Trouble with concentration or focus</li> <li>● Other executive function concerns</li> </ul>	All ages

<p><b>Mental health counseling and other support</b></p>	<p>Emotional and behavioral supports provided by a school psychologist, school social worker, or school counselor. This may involve individual counseling sessions, social skills groups, or consultation with parents and teachers.</p>	<ul style="list-style-type: none"> <li>● Social skills deficits</li> <li>● Emotion management issues</li> <li>● Self-harm</li> <li>● Refusal to go to school</li> <li>● Inattentive behaviors</li> <li>● Identity development</li> <li>● Family systems concerns</li> <li>● Other social, emotional, or behavioral problems</li> </ul>	<p>All ages</p>
<p><b>Specialized instruction or special education</b></p>	<p>Explicit and individualized academic instruction with a licensed special education teacher. Helps students meet academic goals as outlined in an IEP.</p>	<p>A student's qualification as a student with a disability under the laws of IDEA.</p>	<p>All ages</p>
<p><b>Assistive technology</b></p>	<p>Any device or equipment that helps a student function more effectively at school. Can be low tech (such as special pencil grips, a slant board for writing, a handheld magnifier, books on tape) or high tech (such as speech-to-text or text-to-speech software or other communication devices).</p>	<p>A student's inability to access the curriculum without these tools.</p>	<p>All ages</p>



<b>Physical therapy</b>	Therapy to improve strength, balance, coordination, mobility.	Physical disabilities that impact a student's ability to learn and access the curriculum.	All ages
<b>Academic intervention</b>	A student meets in a small group or individually with an interventionist or a classroom teacher for targeted instruction in specific academic skill(s). This intervention may occur daily or weekly.	A student failing to make adequate progress on specific academic skills. Students do not require an IEP to qualify for intervention.	All ages
<b>Accommodations</b>	Slight changes in presentation, format, or response procedures that alter how students learn or demonstrate knowledge of the content. Accommodations do not alter the actual content learned.	A student who can access their grade-level curriculum with slight alterations to the delivery of content.	All ages
<b>Modifications</b>	Fundamental changes to a curriculum or assessment that alter what the student is learning or is expected to learn.	The need for modifications included in a student's IEP or 504 plan.	All ages

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**Section 5 of 6: Educational assessments**

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Educators and developmental experts use assessments to gather data on a student’s current level of functioning, areas of strength, and areas of need to target for intervention. Early childhood testing should be conducted by multiple early developmental experts in a multidisciplinary developmental assessment. School-age assessments should also encompass a variety of student domains.

A thorough evaluation, including standardized assessment tools, will be most useful for students with SCAs, as there are often constellations of developmental concerns that can impact learning. The following tables outline some possible assessment instruments that may be most useful with students with SCAs.

<b>Educational assessments for babies and children birth to age 5</b>		
<b>Area of concern</b>	<b>Common milestones</b>	<b>Possible assessments*</b>
<b>Physical or motor abilities</b>	<b>Baby (newborn to 1 year):</b> Holding up head, sitting, crawling, picking up objects	Peabody Developmental Motor Scales, 2nd edition (PDMS-2)
	<b>Toddler (1 to 3 years):</b> Running, jumping, climbing stairs, tolerating various sensory inputs	Bayley Scales of Infant Toddler Development (Bayley)  SPM Sensory Processing Measure  The Carolina Curriculum for Infants and Toddlers with Special Needs

<p><b>Cognition</b></p>	<p><b>Baby (newborn to 1 year):</b> Paying attention to faces, recognizing faces, curiosity</p> <p><b>Toddler (1 to 3 years):</b> Following simple directions, using objects, problem solving</p> <p><b>Preschool age (3 to 5 years):</b> Early academic skills such as shapes, colors, counting, identifying letters</p>	<p>Bayley</p> <p>Mullen Scales of Early Learning (MSEL)</p> <p>Wechsler Preschool and Primary Scale of Intelligence (WPPSI)</p> <p>Differential Ability Scales (DAS)</p> <p>Woodcock Johnson Tests of Early Cognitive and Academic Development (WJ ECAD)</p>
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<p><b>Communication</b></p>	<p><b>Baby (birth to 1 year):</b> Imitating sounds, responding to name, using simple gestures</p> <p><b>Toddler (1 to 3 years):</b> Using words and sentences, naming objects, expressing emotion</p>	<p>Bayley</p> <p>MSEL</p> <p>Rossetti Infant Toddler Play Scale</p> <p>Preschool language Scale-5</p> <p>Clinical Evaluation of Language Fundamentals Preschool -3 (CELF-Preschool 3)</p> <p>Peabody Picture Vocabulary Test, Fifth Edition (PPVT-5)</p> <p>Expressive Vocabulary Test, Third Edition (EVT-3)</p> <p>The Arizona Articulation Proficiency Scale -Third Revision (Arizona-3): Goldman-Fristoe Test of Articulation- Third Edition (GFTA-3)</p> <p>The Articulation Screener on the PLS-5</p>
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<p><b>Social-emotional development</b></p>	<p><b>Baby (birth to 1 year):</b> Smiling, social anxiety, reacting to caregivers</p> <p><b>Toddler (1 to 3 years):</b> Playing near and with peers, imitating play of others, throwing temper tantrums</p>	<p>Bayley</p> <p>Autism Diagnostic Observation Schedule (ADOS)</p> <p>Behavior Assessment Scales for Children (BASC)</p> <p>Parent &amp; Teacher Rating Forms-Preschool</p>
<p><b>Adaptive skills</b></p>	<p><b>Baby (birth to 1 year):</b> Holding and drinking from a bottle or cup, feeding self</p> <p><b>Toddler (1 to 3 years):</b> Getting dressed, toilet training, basic hygiene skills</p>	<p>Vineland Adaptive Behavior Scales (Vineland)</p> <p>Adaptive Behavior Assessment System (ABAS)</p> <p>Scales of Independent Behavior- Revised (SIB-R)</p>

\*Multidisciplinary teams use a variety of tests to assess areas of need. These are just some examples of tests that can be used to address areas of concern.

<p><b>Educational assessments for elementary school students</b></p>	
<p><b>Area of concern</b></p>	<p><b>Possible assessments*</b></p>

<b>Cognition</b>	<p>Wechsler Intelligence Scale for Children (WISC)</p> <p>Differential Ability Scales (DAS)</p> <p>Woodcock Johnson Tests of Cognitive Abilities (WJ)</p>
<b>Adaptive skills</b>	<p>Vineland Adaptive Behavior Scales (Vineland)</p> <p>Adaptive Behavior Assessment System (ABAS)</p> <p>Scales of Independent Behavior- Revised (SIB-R)</p>
<b>Speech and language skills</b>	<p>Peabody Picture Vocabulary Test, Edition 4 or 5 (PPVT-4/5)</p> <p>Expressive Vocabulary Test, Third Edition (EVT-3)</p> <p>Test for the Reception of Grammar (TROG)</p> <p>The WORD TEST Elementary (ages 6 to 11)</p> <p>Clinical Evaluation of Language Fundamentals-5 (ages 5 to 8 and 9-21)</p> <p>Oral Passage Understanding Scale (OPUS) (ages 5 to 21)</p> <p>The Arizona Articulation Proficiency Scale -Third Revision (Arizona-3): Goldman-Fristoe Test of Articulation- Third Edition (GFTA-3)</p>
<b>Motor skills</b>	<p>Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery-VMI)</p> <p>DayC-2 Developmental Assessment of Young Children, 2nd Ed.</p> <p>PDMS-2 Peabody Developmental Motor Scales</p> <p>The Bruininks-Oseretsky Test of Motor Proficiency, Second Edition (BOT-2)</p> <p>SPM Sensory Processing Measure</p>

<b>Social skills/ Behavior/Mental health</b>	Autism Diagnostic Observation Schedule (ADOS)  Behavior Assessment Scales for Children, (BASC) Parent/ Teachers Rating Forms  Conners  NICHQ Vanderbilt Assessment Scales (Vanderbilt)  Multidimensional Anxiety Scale for Children (MASC)
<b>Executive function</b>	Behavior Rating Inventory of Executive Function (BRIEF)  Conners Kiddie Continuous Performance Task (K-CPT)  NIH Cognitive Toolbox (NIHTB-CB) EF subtests (Flanker, Dimensional Change Card Sort, List Sorting, and Picture Sequence Memory)
<b>Academic skills</b>	Wechsler Individual Achievement Test (WIAT)  Woodcock Johnson Test of Achievement (WJ)  Gray Oral Reading Test (GORT)  Comprehensive Test of Phonological Processing (CTOPP)

\*School districts use a variety of tests to assess areas of need. These are just some examples of tests that can be used to address areas of concern.

<b>Educational assessments middle and high school students</b>	
<b>Area of concern</b>	<b>Possible assessments*</b>

<b>Cognition</b>	<p>Wechsler Intelligence Scale for Children (WISC)</p> <p>Wechsler Adult Intelligence Scale (WAIS)</p> <p>Differential Ability Scales (DAS)</p> <p>Woodcock Johnson Tests of Cognitive Abilities (WJ)</p>
<b>Adaptive skills</b>	<p>Vineland Adaptive Behavior Scales (Vineland)</p> <p>Adaptive Behavior Assessment System (ABAS)</p> <p>Scales of Independent Behavior- Revised (SIB-R)</p>
<b>Speech and language</b>	<p>Peabody Picture Vocabulary Test, Edition 4 or 5 (PPVT-4/5)</p> <p>Expressive Vocabulary Test, third edition (EVT-3)</p> <p>Test for the Reception of Grammar (TROG)</p> <p>The WORD TEST Elementary (ages 6-11) or Adolescent (ages 12-17)</p> <p>Clinical Evaluation of Language Fundamentals 5 Metalinguistics (ages 9-21)</p> <p>Understanding Spoken Paragraphs, formulated Sentences from the Clinical Evaluation of Language Fundamentals-5</p> <p>Oral Passage Understanding Scale (OPUS) (ages 5 to 21)</p>
<b>Motor skills</b>	<p>Beery-Buktenica Developmental Test of Visual-Motor Integration (Beery-VMI)</p>



<p><b>Social skills/ Behavior/Mental health</b></p>	<p>Autism Diagnostic Observation Schedule (ADOS)</p> <p>Behavior Assessment Scales for Children, (BASC) Parent, Teacher, &amp; Self Rating Forms</p> <p>Multidimensional Anxiety Scale for Children (MASC)</p> <p>Reynolds Adolescent Depression Scale (RADS)</p> <p>Conners</p> <p>NICHQ Vanderbilt Assessment Scales (Vanderbilt)</p>
<p><b>Executive function</b></p>	<p>Behavior Rating Inventory of Executive Function (BRIEF)</p> <p>Conners Continuous Performance Task (CPT)</p> <p>NIH Cognitive Toolbox (NIHTB-CB) EF subtests (Flanker, Dimensional Change Card Sort, List Sorting, and Picture Sequence Memory)</p>
<p><b>Academic skills</b></p>	<p>Wechsler Individual Achievement Test (WIAT)</p> <p>Woodcock Johnson Test of Achievement (WJ)</p> <p>Gray Oral Reading Test (GORT)</p> <p>Comprehensive Test of Phonological Processing (CTOPP)</p>

\* School districts use a variety of tests to assess areas of need. These are just some examples of tests that can be used to address areas of concern.

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## Section 6 of 6: Educational strategies

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Students with X and Y chromosome variations present with a wide range of educational strengths and needs. We recommend an individualized approach to educational planning. A team approach is also warranted because development does not happen in a vacuum, but rather across developmental domains.

We separate the developmental domains below for ease of locating specific information, but keep in mind that teams can target multiple domains at one time. For example, when working on gross motor skills with an obstacle course, you can also visually and verbally label the items, actions, and directional concepts to build vocabulary and language skills.

We encourage educational teams to meet on a regular basis, communicate regularly, or both so that all team members, including parents, are consistently addressing the target goal areas. See below for recommended evidence-based educational strategies organized by specific documented areas of risk for students with SCAs.

<b>Recommended evidence-based educational strategies for early childhood (ages birth to 5 years)</b>	
<b>Speech and language</b>	
<b>Receptive language</b>	Create a language rich environment by providing numerous opportunities for communication. Label items and actions, read books, tell oral stories, and sing simple songs.
	Build communication into daily routines (such as mealtimes, bath time, or time in the car) to build vocabulary and to enhance communication.
	Provide simple and short prompts and verbal instructions.
	Talk about what you are doing. Narrate what the child is doing.

<b>Expressive language</b>	Pause before providing the child's desired object or activity to encourage a communicative response (such as pointing, eye contact, or verbalization).
	Imitate the child's verbalizations as if you are carrying on a conversation.
	Repeat and extend the child's verbalizations. (Child: "Ball!" Adult: "Ball! Yes, I throw the ball!").

<b>Motor skills</b>	
<b>Fine motor skills</b>	Provide the child with a variety of sensory materials (such as sand, playdough, or shaving cream) to explore.
	Ask the child to find hidden objects. Place desirable materials into see-through containers to encourage the child to use both hands to retrieve objects.
<b>Gross motor skills</b>	Encourage the child to complete simple obstacle courses (such as crawling through a tunnel or standing up and then picking up a ball).

<b>Early academics</b>	
<b>Pre-reading</b>	Play rhyming games, sing songs, recite nursery rhymes, and read books with rhyming passages. Repeating the same songs and books is beneficial.
	Play simple games such as "I Spy" or "Guess what I'm thinking of" or "Go find it!" using first letter sounds (for example, "I'm thinking of something that starts with ssssss.>").
	Play word games such as breaking words apart ("Say cowboy. Now say cowboy without boy.") or putting sounds together ("Say /p/. Now say /ig/. Put them together and say P-ig! Pig!"). Use visual images to encourage motivation.

<b>Early math</b>	Play sorting games with motivating manipulatives (such as sorting kids by shirt colors, sorting jelly beans for a treat, or sorting different colored hearts on Valentine’s day).
	Build with blocks to teach counting, symmetry, patterns.

<b>Social emotional</b>	
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<b>Self-management</b>	Provide a cozy corner or other quiet spot in the classroom where children can take a break and practice self-regulation. Include visual images of emotion faces and visual cues for self-management strategies (such as stop and think, deep breathing, counting) as well as bubble timers, fidget toys, books, stuffed animals, weighted blankets, and coloring supplies.
	Provide choices throughout the day whenever appropriate.
	Help children to recognize and label emotions. Use photographs of emotion faces, label emotions of characters in stories, and label your own emotions throughout the day as a model.
	Teach explicit steps for calming down (such as “Stop, take a deep breath, count to 5” or the “Turtle Technique” from CSEFEL).

<b>Social skills</b>	Use social stories (brief descriptive stories with images) to provide behavioral expectations for routine or novel social situations (such as how to participate in circle time, ways to play at recess, or how to invite a friend to play).
	Recognize and provide positive feedback when you observe prosocial behaviors. Consider creating a class-wide positive behavior system such as a “super friend” or “helper of the day” award system for the entire preschool class.

Teach, role play, and practice social problem-solving skills (such as taking turns, sharing, asking for help, walking away, and compromising).

## Recommended evidence-based educational strategies for school-aged children (grades K to 12)

### Academic

#### Reading decoding

Use text-to-voice software to assist the student with decoding difficult words.

For early grade students, provide direct and systematic instruction in phonics. A whole language approach to reading may make reading acquisition more difficult for a student with an SCA.

#### Reading comprehension

Consider using multiple choice, fill-in-the-blank, or true/false questions to assess comprehension, rather than open-ended questions.

Use a note-taking strategy to mark important ideas in the text during reading. This can include highlighting, making notes or pictures in the margins, or completing a graphic organizer during reading.

Provide audio books or an e-reader.

Read a summary or a simplified text about a topic prior to the assigned reading to provide background knowledge and vocabulary.

Provide chunking of texts with periodic “check-ins” to assess for understanding throughout the reading.

#### Writing

Use voice-to-text software and have the student go back over the text to edit for spelling and grammar errors.

	Use word-prediction software to reduce the number of keystrokes required.
	Consider modifying tests that require written responses, or providing alternate options for demonstrating knowledge (such as oral presentations or take-home essays).
	Provide teacher notes ahead of lessons and a scribe or peer notetaker if the student takes too long to produce writing in the classroom or a testing situation.
	Provide templates, outlines, and graphic organizers so the student can chunk written work into manageable segments and receive frequent feedback on elements of writing.
	Use a “smart pen” (such as the LiveScribe pen) so the student can digitize handwritten class notes.
<b>Mathematics</b>	Provide multisensory and systematic math instruction with concrete, structured materials the student can manipulate while problem solving.
	Allow the student to use a calculator whenever possible so that inefficient fact recall does not interfere with their ability to move forward in math procedural knowledge.

<b>Executive function</b>	
<b>Inattention</b>	Provide multiple sources for the student to get instructions (such as written instructions on a whiteboard, verbal explanation, a virtual classroom, and a handout with a detailed rubric)
	Provide nonverbal cues for the student.
	Encourage self-monitoring strategies (such as wearable devices, reminders on a cell phone, or a timer on desk to prompt the student to refocus at predetermined time intervals).

	Provide frequent check-ins for understanding.
<b>Organization and time management</b>	Provide time limits for task completion during independent work time.
	Schedule routine (daily or weekly) adult support to help the student organize their binder, desk, and locker.
	Provide direct instruction in, and frequent monitoring of, the student’s personal planner system. Try a variety of planners (such as digital and color-coded planners) to determine the best fit for the individual student.
	Chunk assignments in small parts, with frequent teacher check-ins and reinforcement.
	Encourage the student to create and use checklists.
<b>Transitioning/Cognitive shifting</b>	Provide verbal and nonverbal (visual or sound) cues to prepare the student for upcoming transitions.
	Provide a transition object or task to help the student move from one activity to the next.
	Allow extra time for the student to transition to the next activity.

<b>Social emotional behavioral</b>	
<b>Anxiety</b>	Help the student to identify triggers for anxiety in the classroom, cafeteria, or playground.
	Teach diaphragmatic breathing, progressive muscle relaxation, and positive self-talk messages the student can use when stressed.
	Provide “pressure passes” or “take a break” cards for the student to leave class as needed for self-regulation.

	<p>Provide a quiet space in the classroom, main office, counselor’s office, or hallway with visual cues for calm-down strategies as well as a relaxation tool kit with supplies (such as bubble timers, thinking putty, meditation apps on an iPad, coloring pages, comic books or magazines, stress balls, or snacks).</p>
<b>Mood/Depression</b>	<p>Monitor the student closely for social withdrawal, negative affect, and loss of pleasure. Refer the student to mental health providers for a check-in as needed.</p>
	<p>Help the student to identify their strengths and provide opportunities for the student to showcase those strengths in the classroom.</p>
	<p>Provide a back-and-forth notebook for the student and teacher to communicate about the student’s emotional needs privately.</p>
	<p>Validate, and do not dismiss, the student’s emotions. Even if they seem inappropriate in the context to you, the emotions are very real for the child. Students require empathy to help them self-regulate.</p>
	<p>Take any indications of suicidal ideation seriously and report them to the student’s family and mental health providers immediately.</p>
<b>Social skills</b>	<p>Provide direct instruction for social-emotional learning in small group or one-on-one settings. Focus on social thinking skills, empathy, steps for social problem solving, and self-advocacy.</p>
	<p>Hold classroom meetings to discuss social problems in the classroom. Engage students in overt social problem-solving activities with teacher guidance.</p>
	<p>Assign, train, and reinforce a team of students (such as a circle of friends program) who will support and partner with the student of concern in social settings (such as the lunch room, recess, partner activities, free-time, and field trips).</p>



	Engage the student in service-learning activities.
<b>Externalizing behaviors/ Hyperactivity</b>	Create daily report cards with goals and reinforcement for prosocial behaviors.
	Create an individualized behavior plan that identifies the underlying function of the student's behavior and find a prosocial way for the student to address this need.
	Schedule frequent motor breaks for the student. Allow the student to move in the classroom, walk to another area of the school, or find other ways to get movement.
	Post visual reminders or cues for behavior expectations in each setting of the school.
	Use planned ignoring for behaviors that are annoying and attention-seeking, but are not unsafe or disruptive to classmates.
	Use collaborative problem-solving techniques with the student and teacher to identify behavior problems and brainstorm possible solutions.

<b>Speech and language</b>	
<b>Receptive language</b>	Provide speech language therapy consultation in the classroom to help strategize ways to support the student's language skills throughout the school day. This may include a visual schedule of class activities, which can increase participation and decrease anxiety.
	Review and practice vocabulary associated with each new lesson in the curriculum.
	Provide support for reading and writing.

	Teach basic language concepts to improve the student's understanding and participation throughout the school day.
<b>Expressive language</b>	Ensure consultation and collaboration with the classroom teacher and the IEP team to adequately address expressive communication skills needed for the student to participate throughout the school day.
	Focus on vocabulary, language concepts, and grammatical structure. Some ideas include: Playing word games such as naming items in a category at the rate of one per second (such as furniture, animals, breakfast foods), completing riddles based on descriptions of objects (such as, "I'm thinking of an animal that lives on a farm and says moo. What is it?"), or completing sentences (such as, "You drive a? A banana is a? You sit on a? You eat soup with a?").
	Expand on narrative skills. You can do this through sharing picture books, practicing sequencing events, and retelling stories.
<b>Social pragmatics</b>	Encourage collaboration among the counselor, teacher, and other team members to ensure consistency with addressing social skills across settings.
	Explicitly teach social skills appropriate to the student's age and developmental level. This may include skills such as discourse skills, perspective taking, problem solving, play skills, and social emotional regulation.
	Provide opportunities to practice social skills in small groups (such as a "lunch bunch").

<b>Motor</b>	
<b>Fine motor</b>	Encourage the student to use small broken crayons to draw or make shapes. This will help to increase a 3-finger grasp.

	Use a variety of materials to create prewriting shapes (such as lines, curves, circles, and squares).
	Provide vestibular and proprioceptive opportunities (such as balancing on a large ball, swinging, crawling through tunnels, sitting in a “large sock,” or playing in a box or under a table).
	Communicate with an occupational therapist for specific fine-motor and sensory activities.
	Make sure the student’s seating is ergonomically correct and that the student has their feet on the floor during tabletop tasks.
<b>Gross motor skills</b>	Demonstrate a variety of postures during yoga (movement breaks).
	Give the student opportunities to help while using both hands to push heavy containers filled with toys or books. The student can also pushchairs to different areas of the room.
<b>Activities of daily living</b>	Encourage the student to place their backpack on a hook, open their own snack container, wash their hands, and do other basic daily tasks independently.
	Explicitly teach daily living skills from a young age. Use visuals to sequence activities and promote independence.

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**Note:** *This guideline was authored by Erin Frith and Talia Thompson, PhD. It has been approved by and represents the current consensus of the members of the AXYS Clinical & Research Consortium.*

**The AXYS Clinical & Research Consortium was founded in 2015 and exists to:**

- **Make life easier for those seeking evaluation and treatment.**
- **Bring consistency to treatment that is consensus and/or evidence-based.**
- **Advance the overall X&Y variation field through coordinated efforts including research.**
- **Bring clinical excellence to the field of X&Y variations.**



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