

**Subject:** Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

Guideline #:CG-BEH-01Publish Date:09/20/2018Status:ReviewedLast Review Date:02/27/2018

## **Description**

This document addresses various tools used in the screening and testing of individuals with suspected Autism Spectrum Disorders (ASDs) and Rett syndrome. ASDs, as defined in the fifth edition of the American Psychiatric Association's (APA) *Diagnostic and Statistical Manual of Mental Disorders* (DSM-5), include disorders previously referred to as:

- Atypical autism
- Asperger's disorder
- Childhood autism
- Childhood disintegrative disorder
- Early infantile autism
- High-functioning autism
- Kanner's autism
- Pervasive developmental disorder not otherwise specified

NOTE: Please see the following related documents for additional information:

- CG-BEH-02 Adaptive Behavioral Treatment for Autism Spectrum Disorder
- CG-BEH-15 Activity Therapy for Autism Spectrum Disorders and Rett Syndrome
- CG-MED-75 Medical and Other Non-Behavioral Health Related Treatments for Autism Spectrum Disorders and Rett syndrome
- CG-MED-76 Magnetic Source Imaging and Magnetoencephalography
- CG-MED-77 SPECT/CT Fusion Imaging
- DRUG.00003 Chelation Therapy
- GENE.00021 Chromosomal Microarray Analysis (CMA) for Developmental Delay, Autism Spectrum Disorder, Intellectual Disability (Intellectual Developmental Disorder) and Congenital Anomalies
- RAD.00002 Positron Emission Tomography (PET) and PET/CT Fusion
- RAD.00023 Single Photon Emission Computed Tomography (SPECT) Scans for Noncardiovascular Indications
- RAD.00044 Magnetic Resonance Neurography

## **Clinical Indications**

#### **Medically Necessary:**

Assessment for ASDs and Rett syndrome is considered **medically necessary** when **any** of the following criteria A through C **and both** criteria D and E have been met:

A. Presence of any of the following signs or symptoms in infants or young children:

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

## Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

- 1. No babbling by 12 months; or
- 2. No gesturing (for example, pointing, waving bye-bye) by 12 months; or
- 3. No single words by 16 months; or
- 4. No 2-word spontaneous (not just echolalic) phrases by 24 months; or
- 5. Any loss of any language or social skills at any age; or
- 6. None-to-little mutual gaze or joint attention;

or

- B. Persistent deficits in social communication and social interaction across multiple contexts, as manifested by **any** of the following:
  - 1. Deficits in social-emotional reciprocity; or
  - 2. Deficits in nonverbal communicative behaviors used for social interaction; or
  - 3. Deficits in developing, maintaining and understanding relationships;

or

- C. Restricted, repetitive patterns of behavior, interests, or activities as manifested by the following
  - 1. Stereotyped or repetitive motor movements, use of objects, or speech; or
  - 2. Insistence on sameness, inflexible adherence to routines, or ritualized patterns of verbal or nonverbal behavior; **or**
  - 3. Highly restricted, fixated interests that are abnormal in intensity or focus; or
  - 4. Hyper- or hypoactivity to sensory inputs or unusual interest in sensory aspects of the environment;

#### and

D. Symptoms must be present in the early developmental period (but may not become fully manifest until social demands exceed limited capacities, or may be masked by learning strategies in later life);

#### and

E. Symptoms cause clinically significant impairment in social, occupational, or other important areas of current functioning.

Note: The following services may be included in the assessment of the individuals with suspected ASDs and Rett syndrome:

- A. Medical evaluation (complete medical history and physical examination).
- B. Evaluation by speech-language pathologist.
- C. Parent and/or child interview (including siblings of children with ASDs).
- D. Audiological hearing evaluation including frequency-specific brainstem auditory evoked response.
- E. Measurement of blood lead level if the child exhibits developmental delay and pica, or lives in a high-risk environment. Additional periodic lead screening can be considered if the pica persists.
- F. Genetic counseling for parents of a child with ASDs.
- G. Genetic testing, specifically high resolution chromosome analysis (karyotype) and DNA analysis for fragile X syndrome in the presence of intellectual developmental disorder (or if intellectual developmental disorder cannot be excluded) if there is a family history of fragile X or intellectual developmental disorder of undetermined etiology, or if dysmorphic features are present.
- H. Neuropsychological evaluation including developmental testing in the areas of social skills, intellect, adaptation and an ASD specific screening tool.
- I. Selective metabolic testing if the child exhibits **any** of the following:

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

# Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

- 1. Clinical and physical findings suggestive of a metabolic disorder (for example, lethargy, cyclic vomiting, or early seizure); **or**
- 2. Dysmorphic or coarse features; or
- 3. Evidence of intellectual developmental disorder; or
- 4. Intellectual developmental disorder cannot be ruled out; or
- 5. Occurrence or adequacy of newborn screening for a birth defect is questionable; or
- 6. History of developmental regression.
- J. Sleep-deprived EEG study only if the child exhibits any of the following conditions:
  - 1. Clinical seizures; or
  - 2. High suspicion of subclinical seizures; or
  - 3. Symptoms of developmental regression (clinically significant loss of social and communicative function) at any age, but especially in toddlers and pre-schoolers.

#### **Not Medically Necessary:**

The following tests/tools are considered **not medically necessary** for the assessment of ASDs and Rett syndrome:

- A. Allergy testing (including but not limited to food allergy for gluten, casein, Candida, and other molds).
- B. Erythrocyte glutathione peroxides studies.
- C. Event-related brain potentials (other than frequency-specific brainstem auditory evoked response, as noted above).
- D. Hair analysis for trace elements.
- E. Intestinal permeability studies.
- F. Stool analysis.
- G. Tests for celiac antibodies.
- H. Tests for immunologic or neurochemical abnormalities.
- I. Tests for micronutrients such as vitamin levels.
- J. Tests for mitochondrial disorders including lactate and pyruvate.
- K. Tests for urinary peptides.

## **Coding**

The following codes for treatments and procedures applicable to this document are included below for informational purposes. Inclusion or exclusion of a procedure, diagnosis or device code(s) does not constitute or imply member coverage or provider reimbursement policy. Please refer to the member's contract benefits in effect at the time of service to determine coverage or non-coverage of these services as it applies to an individual member.

CPT	
	Codes include, but are not limited to, the following:
81243	FMR1 (Fragile X mental retardation 1) (eg, fragile X mental retardation) gene analysis;
	evaluation to detect abnormal (eg, expanded) alleles
81244	FMR1 (Fragile X mental retardation 1) (eg, fragile X mental retardation) gene analysis;
	characterization of alleles (eg, expanded size and methylation status)
81401	Molecular pathology procedure, Level 2 (eg, 2-10 SNPs, 1 methylated variant, or 1
	somatic variant [typically using nonsequencing target variant analysis] or detection of a
	dynamic mutation disorder/triplet repeat) [when specified as the following]:

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

# Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

	• AFF2 (AF4/FMR2 family, member 2 [FMR2]) (eg, fragile X mental retardation 2
01.40.4	[FRAXE]), evaluation to detect abnormal (eg, expanded) alleles
81404	Molecular pathology procedure, Level 5 (eg, analysis of 2-5 exons by DNA sequence
	analysis, mutation scanning or duplication/deletion variants of 6-10 exons, or
	characterization of a dynamic mutation disorder/triplet repeat by Southern blot analysis)
	[when specified as the following]:
	• AFF2 (AF4/FMR2 family, member 2 [FMR2]) (eg, fragile X mental retardation 2
	[FRAXE]), characterization of alleles (eg, expanded size and methylation status);
	• <i>NLGN4X</i> ( <i>neuroligin 4</i> , <i>X-linked</i> ) (eg, autism spectrum disorders),
	duplication/deletion analysis
81405	Molecular pathology procedure, Level 6 (eg, analysis of 6-10 exons by DNA sequence
	analysis, mutation scanning or duplication/deletion variants of 11-25 exons, regionally
	targeted cytogenomic array analysis) [when specified as the following]:
	• <i>NLGN3</i> ( <i>neuroligin 3</i> ) (eg, autism spectrum disorders), full gene sequence;
	• <i>NLGN4X</i> ( <i>neuroligin 4</i> , <i>X-linked</i> ) (eg, autism spectrum disorders), full gene sequence
83655	Lead testing
88245-88264	Chromosome analysis [includes codes 88245, 88248, 88249, 88261, 88262, 88263,
	88264]
88280-88289	Chromosome analysis [includes codes 88280, 88283, 88285, 88289]
92521	Evaluation of speech fluency (eg, stuttering, cluttering)
92522	Evaluation of speech sound production (eg, articulation, phonological process, apraxia,
	dysarthria);
92523	Evaluation of speech sound production (eg, articulation, phonological process, apraxia,
	dysarthria); with evaluation of language comprehension and expression (eg, receptive and
00704	expressive language)
92524	Behavioral and qualitative analysis of voice and resonance
92550, 92552-	Audiologic function tests with medical diagnostic evaluation [includes codes 92550,
92588	92552, 92553, 92555, 92556, 92557, 92558, 92559, 92560, 92561, 92562, 92563, 92564,
	92565, 92567, 92568, 92570, 92571, 92572, 92575, 92576, 92577, 92579, 92582, 92583,
0.501.6	92584, 92585, 92586, 92587, 92588]
95816	Electroencephalogram (EEG); including recording awake and drowsy
96040	Medical genetics and genetic counseling services, each 30 minutes face-to-face with
06101	patient/family
96101	Psychological testing (includes psychodiagnostic assessment of emotionality, intellectual
	abilities, personality and psychopathology, eg, MMPI, Rorschach, WAIS), per hour of the
	psychologist's or physician's time, both face-to-face time administering tests to the patient
0.5100	and time interpreting these test results and preparing the report
96102	Psychological testing (includes psychodiagnostic assessment of emotionality, intellectual
	abilities, personality and psychopathology, eg, MMPI and WAIS), with qualified health
	care professional interpretation and report, administered by technician, per hour of
0.6102	technician time, face-to-face
96103	Psychological testing (includes psychodiagnostic assessment of emotionality, intellectual
	abilities, personality and psychopathology, eg, MMPI), administered by a computer, with
	qualified health care professional interpretation and report

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

## Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

	1
96105	Assessment of aphasia (includes assessment of expressive and receptive speech and
	language function, language comprehension, speech production ability, reading, spelling,
	writing, eg, by Boston Diagnostic Aphasia Examination) with interpretation and report,
	per hour
96110	Developmental screening, (eg, developmental milestone survey, speech and language
	delay screen), with scoring and documentation, per standardized instrument
96111	Developmental testing, (includes assessment of motor, language, social, adaptive, and/or
	cognitive functioning by standardized developmental instruments) with interpretation and
	report
96116	Neurobehavioral status exam (clinical assessment of thinking, reasoning and judgment,
	eg, acquired knowledge, attention, language, memory, planning and problem solving, and
	visual spatial abilities), per hour of the psychologist's or physician's time, both face-to-
0.6110	face time with the patient and time interpreting test results and preparing the report
96118	Neuropsychological testing (eg, Halstead-Reitan Neuropsychological Battery, Wechsler
	Memory Scales and Wisconsin Card Sorting Test), per hour of the psychologist's or
	physician's time, both face-to-face time administering tests to the patient and time
96119	interpreting these test results and preparing the report Neuropsychological testing (eg, Halstead-Reitan Neuropsychological Battery, Wechsler
90119	Memory Scales and Wisconsin Card Sorting Test), with qualified health care professional
	interpretation and report, administered by technician, per hour of technician time, face-to-
	face
96120	Neuropsychological testing (eg, Wisconsin Card Sorting Test), administered by a
J0120	computer, with qualified health care professional interpretation and report
99201-99215	Evaluation and Management services [includes codes 99201, 99202, 99203, 99204,
	99205, 99211, 99212, 99213, 99214, 99215]
HCPCS	
	Codes include, but are not limited to, the following:
G0451	Development testing, with interpretation and report, per standardized instrument form
S0265	Genetic counseling, under physician supervision, each 15 minutes
S9152	Speech therapy, re-evaluation
ICD-10 Diagnosis	
F84.0	Autistic disorder
F84.2	Rett's syndrome
F84.3	Other childhood disintegrative disorder
F84.5	Asperger's syndrome
F84.8	Other pervasive developmental disorders
F84.9	Pervasive developmental disorder, unspecified
Z13.40-Z13.49	Encounter for screening for certain developmental disorders in childhood

#### **Discussion/General Information**

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

## Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

Systematic screening for conditions like autism spectrum disorder is an important part of managing the health of children and adolescents. As with many conditions, the early identification of ASDs and Rett syndrome allow for early intervention, which has been shown to have a significant positive impact on treatment outcomes.

In May 2013, the American Psychiatric Association (APA) released the 5<sup>th</sup> edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5). This edition of the DSM includes several significant changes over the previous edition, including combining several previously separate diagnoses under the single diagnosis of "autism spectrum disorder" (ASD). This diagnosis included the following disorders, previously referred to as: atypical autism, Asperger's disorder, childhood autism, childhood disintegrative disorder, early infantile autism, high-functioning autism, Kanner's autism, and pervasive developmental disorder not otherwise specified. All of these conditions are now considered under one diagnosis, ASD. It should be noted that Rett syndrome is not included in the new DSM-5 ASD diagnostic group but is included in this discussion due to similar condition presentations and diagnostic work up.

The DSM-5 describes the essential diagnostic features of autism spectrum disorders as both a persistent impairment in reciprocal social communication and restricted and repetitive patterns of behavior, interest or activities. These attributes are present from early childhood and limit or impair everyday functioning. Parents may note symptoms as early as infancy, and the typical age of onset is before 3 years of age. Symptoms may include problems with using and understanding language; difficulty relating to or reciprocating with people, objects, and events; lack of mutual gaze or inability to attend events conjointly; unusual play with toys and other objects; difficulty with changes in routine or familiar surroundings, and repetitive body movements or behavior patterns. Children with childhood disintegrative disorder are an exception to this description, in that they exhibit normal development for approximately 2 years followed by a marked regression in multiple areas of function.

Children with ASD vary widely in abilities, intelligence, and behaviors. Some children do not speak at all, others speak in limited phrases or conversations, and some have relatively normal language development. Repetitive play skills, resistance to change in routine and inability to share experiences with others, and limited social and motor skills are generally evident. Unusual responses to sensory information, such as loud noises and lights, are also common. Children unaffected by ASDs can exhibit unusual behaviors occasionally or seem shy around others sometimes without having ASD. What sets children with ASD apart is the consistency of their unusual behaviors. Symptoms of the disorder have to be present in all settings, not just at home or at school, and over considerable periods of time. With ASD, there is a lack of social interaction, impairment in nonverbal behaviors, and a failure to develop normal peer relations. A child with an ASD tends to ignore facial expressions and may not look at others; other children may fail to respect interpersonal boundaries and come too close and stare fixedly at another person.

The exact causes of autism are unknown, although genetic factors are strongly implicated. A study released by the Center for Disease Control and Prevention (2014) indicates that the incidence of ASD was as high as 1 in 68.

In 2014, the American Academy of Child and Adolescent Psychiatry (AACAP) published their practice parameter for the assessment and treatment of children and adolescents with ASDs. They made the following recommendations for the assessment of children with ASDs:

• <u>Recommendation 1</u>. The developmental assessment of young children and the psychiatric assessment of all children should routinely include questions about ASD symptomatology [CS].

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

## Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

- Recommendation 2. If the screening indicates significant ASD symptomatology, a thorough diagnostic evaluation should be performed to determine the presence of ASD [CS].
- Recommendation 3. Clinicians should coordinate an appropriate multidisciplinary assessment of children with ASD [CS].

These recommendations are based on "clinical standard" or "CS", which they state is applied to recommendations based on rigorous empirical evidence and/or overwhelming clinical consensus.

The American Academy of Pediatrics published recommendations for early screening for ASDs (Zwaigenbaum, 2015b). Their first recommendation states, "Evidence supports the usefulness of ASD-specific screening at 18 and 24 months. ASD screening before 24 months may be associated with higher false-positive rates than screening at 24 months but may still be informative."

Finally, the U.S. Preventive Services Task Force published their recommendations for ASD screening in young children in 2016. Their recommendation for children ages 18 to 30 months stated:

The USPSTF concludes that the current evidence is insufficient to assess the balance of benefits and harms of screening for autism spectrum disorder (ASD) in young children for whom no concerns of ASD have been raised by their parents or a clinician.

This recommendation is provided with an "I" grade, which indicates that "the USPSTF concluded that the current evidence is insufficient to assess the balance of benefits and harms of the service. Evidence is lacking, of poor quality, or conflicting, and the balance of benefits and harms cannot be determined."

The specific DSM-5 diagnostic criteria for ASD are provided below:

## DSM-5 Criteria for Autism Spectrum Disorder\*

#### Diagnostic criteria

- A. Persistent deficits in social communication and social interaction across multiple contexts, as manifested by the following, currently or by history (examples are illustrative, not exhaustive; see DSM- 5 text);
  - 1. Deficits in social-emotional reciprocity, ranging, for example, from abnormal social approach and failure of normal back-and-forth conversation; to reduced sharing of interests, emotions, or affect; to failure to initiate or respond to social interactions.
  - 2. Deficits in nonverbal communicative behaviors used for social interaction, ranging, for example, from poorly integrated verbal and nonverbal communication, to abnormalities in eye contact and body language or deficits in understanding and use of gestures; to a total lack of facial expressions and nonverbal communications.
  - 3. Deficits in developing, maintaining and understanding relationships, ranging, for example, from difficulties adjusting behavior to suit various social context; to difficulties in sharing imaginative play or in making friends; to absence of interest in peers.

Specify current severity:

Severity is based on social communication impairments and restricted, repetitive patterns of behavior (See table 2).

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

# Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

- B. Restricted, repetitive patterns of behavior, interests, or activities as manifested by at least two of the following, currently or by history (examples are illustrative, not exhaustive; see text);
  - 1. Stereotyped or repetitive motor movements, use of objects, or speech (e.g., simple motor stereotypes, lining up toys or flipping objects, echolalia, idiosyncratic phrases).
  - 2. Insistence on sameness, inflexible adherence to routines, or ritualized patterns of verbal or nonverbal behavior (e.g., extreme distress at small changes, difficulties with transitions, rigid thinking patterns, greeting rituals, need to take same route or eat same food every day).
  - 3. Highly restricted, fixated interests that are abnormal in intensity or focus (e.g., strong attachment to or preoccupation with unusual objects, excessively circumscribed or perseverative interests.
  - 4. Hyper- or hypoactivity to sensory inputs or unusual interest in sensory aspects of the environment (e.g., apparent indifference to pain/temperature, adverse response to specific sounds or textures, excessive smelling or touching or objects, visual fascination with lights or movement).

*Specify* current severity:

Severity is based on social communication impairments and restricted, repetitive patterns of behavior (See table 2).

- C. Symptoms must be present in the early developmental period (but may not become fully manifest until social demands exceed limited capacities, or may be masked by learning strategies in later life).
- D. Symptoms cause clinically significant impairment in social, occupational, or other important areas of current functioning.
- E. These disturbances are not better explained by intellectual disability (intellectual developmental disorder) or global developmental delay. Intellectual disability and autism spectrum disorder frequently co-occur; to make comorbid diagnoses of autism spectrum disorder and intellectual disability, social communication should be below that expected for general developmental level.

**Note:** Individuals with a well-established DSM-IV diagnosis of autistic disorder, Asperger's disorder, or pervasive developmental disorder not otherwise specified should be given the diagnosis of autism spectrum disorder. Individuals who have marked deficits in social communication, but whose symptoms do not otherwise meet criteria for autism spectrum disorder, should be evaluated for social (pragmatic) communication disorder. Specify current severity:

With or without accompanying intellectual impairment

With or without accompanying language impairment

Associated with a known medical or genetic condition or environmental factor (Coding note: Use additional code to identify the associated medical or genetic condition)

Associated with another neurodevelopmental, mental, or behavioral disorder (Coding note: Use additional code[s] to identify the associated neurodevelopmental, mental, or behavioral disorder)

With catatonia (refer to the criteria for catatonia associated with another mental disorder, pp. 119-120 for definition). (Coding note: Use additional code 293.89 [F06.1] catatonia associated with autism spectrum disorder to indicate the presence of the comorbid catatonia)

\* From: Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition. DSM-5. American Psychiatric Association. Washington, DC. May 2013. Page 50-51.

Table 2 Severity levels for autism spectrum disorders*		
Severity Level	Social Communication	Restricted, repetitive behaviors

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

Level 3	Severe deficits in verbal and	Inflexibility of behavior, extreme
"Requiring very substantial	nonverbal social communications	difficulty coping with change, or
support"	skills cause severe impairments in	other restricted / repetitive
	functioning, very limited initiation	behaviors markedly interfere with
	of social interactions, and minimal	functioning in all spheres. Great
	response to social overtures from	distress / difficulty changing focus
	others. For example, a person with	or action.
	few words of intelligible speech	
	who rarely initiates interaction and,	
	when he or she does, makes unusual	
	approaches to meet needs only and	
	responds to only very direct social	
	approaches.	
Level 2	Marked deficits in verbal and	Inflexibility of behavior, difficulty
"Requiring substantial support"	nonverbal communication skills;	coping with change, or other
	social impairments apparent even	restricted / repetitive behaviors
	with supports in place; limited	appear frequently enough to be
	initiation of social interactions; and	obvious to the casual observer in a
	reduced or abnormal responses from	variety of context. Distress and or
	others. For example, a person who	difficulty changing focus or action.
	speaks simple sentences, whose	
	interaction is limited to narrow	
	special interests, and who has	
	markedly odd nonverbal	
	communication.	
Level 1	Without supports in place, deficits	Inflexibility of behavior cases
"Requiring support"	in social communication cause	significant interference with
	noticeable impairments. Difficulty	functioning in one or more context.
	initiating social interactions, and	Difficulty switching between
	clear examples of atypical or	activities. Problems of organization
	unsuccessful responses to social	and planning hamper independence.
	overtures of others. May appear to	
	have decreased interest in social	
	interactions. For example, a person	
	who is able to speak in full	
	sentences and engages in	
	communications but whose to-and-	
	fro conversation with others fails,	
	and whose attempts to make friends	
	are odd and typically unsuccessful.	

<sup>\*</sup> From: Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition. DSM-5. American Psychiatric Association. Washington, DC. May 2013. Page 52.

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

## Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

Rett syndrome is a disorder of the nervous system that leads to regression in development, especially in the areas of expressive language and hand use. In most cases, it is caused by a genetic mutation. It occurs almost exclusively in girls and may be misdiagnosed as autism or cerebral palsy.

Seventy-five percent of Rett syndrome cases have been linked to a specific genetic mutation on the X chromosome. This gene contains instructions for creating methyl-CpG-binding protein 2 (MeCP2), which regulates the manufacture of various other proteins. Mutations in the MeCP2 gene cause these other proteins to be produced incorrectly, which damage the maturing brain. Most cases of the mutation arise spontaneously without any traceable cause. However, there also seem to be some clusters within families and certain geographic regions, for example Norway, Sweden, and Northern Italy.

A child affected with Rett syndrome normally follows a standard developmental path for the first 5 months of life. After that time development in communication skills and motor movement in the hands seems to stagnate or regress. After a short period, stereotyped hand movements, gait disturbances, and slowing of the rate of head growth become apparent. Other problems may also be associated with Rett syndrome, including seizures, disorganized breathing patterns while awake and apraxia/dyspraxia (the inability to program the body to perform motor movements). Apraxia/dyspraxia is a key symptom of Rett syndrome and it results in significant functional impairment, interfering with body movement, including eye gaze and speech.

#### **Definitions**

Asperger's Syndrome: A developmental disorder that affects the parts of the brain that control social interaction and communications.

Assessment instruments: Specialized and standardized diagnostic test used to evaluate an individual's performance in specific areas of functioning such as those recommended in the guidelines of the AAP, AAN and the AACAP (for example, learning and communications skills, social interaction, etc.). Examples of this type of instrument include Verbal Behavior Milestones Assessment and Placement Program (VB-MAPP), the Vineland Adaptive Behavior Scale, the Autism Diagnostic Interview-Revised (ADI-R), the Gilliam Autism Rating Scale - Second Edition (GARS-2), etc.

Autism Spectrum Disorders: A collection of associated developmental disorders that affect the parts of the brain that control social interaction and verbal and non-verbal communication.

Childhood Disintegrative Disorder: A developmental disorder characterized by marked regression in multiple areas of functioning following a period of at least 2 years of apparently normal development.

Dysmorphic: A characteristic that is abnormally formed.

Echiolaic: A symptom of some medical conditions characterized by an individual repeating things they hear.

Pica: A symptom of some medical conditions characterized by eating earth, clay or chalk.

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

Page 11 of 13

## **Clinical UM Guideline**

# Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

Rett syndrome: A developmental disorder that affects the parts of the brain that control social interaction, communications, and motor function.

#### References

#### **Peer Reviewed Publications:**

- 1. Charman T, Baird G, Simonoff E, et al. Efficacy of three screening instruments in the identification of autistic-spectrum disorders. Br J Psychiatry. 2007; 191:554-559.
- 2. Witwer AN, Lecavalier L. Autism screening tools: an evaluation of the Social Communication Questionnaire and the Developmental Behaviour Checklist-Autism Screening Algorithm. J Intellect Dev Disabil. 2007; 32(3):179-187.

#### Government Agency, Medical Society, and Other Authoritative Publications:

- 1. American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition. DSM-5. Washington, DC. May 2013.
- 2. Centers for Disease Control and Prevention. Prevalence of Autism Spectrum Disorder Among Children Aged 8 Years Autism and Developmental Disabilities Monitoring Network, 11 Sites, United States, 2010. Morbidity and Mortality Weekly Report (MMWR). 2014; 3(SS02):1-21.
- 3. Filipek PS, Accardo PJ, Ashwal S, et al. American Academy of Neurology and the Child Neurology Society. Practice parameter: screening and diagnosis of autism: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Child Neurology Society. Neurology. 2000; 55(4):468-479. Guideline-reaffirmed 07/28/2006.
- 4. Miller DT, Adam MP, Aradhya S, et al. Consensus statement: chromosomal microarray is a first-tier clinical diagnostic test for individuals with developmental disabilities or congenital anomalies. Am J Hum Genet. 2010; 86(5):749-764.
- 5. Shen T, Dies KA, Holm IA, et al.; Autism Consortium Clinical Genetics/DNA Diagnostics Collaboration. Clinical genetic testing for patients with autism spectrum disorders. Pediatrics. 2010; 125(4):e727-e735.
- 6. Shevell M, Ashwal S, Donley D, et al. Practice parameter: evaluation of the child with global developmental delay: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology. 2003; 60(3):367-380. Guideline reaffirmed 10/15/2005.
- 7. U.S. Preventive Services Task Force (USPSTF): Autism Spectrum Disorder in young children: a systematic evidence review for the U.S Preventive Services Task Force. February 2016. Available at: <a href="https://www.uspreventiveservicestaskforce.org/Page/Document/UpdateSummaryFinal/autism-spectrum-disorder-in-young-children-screening">https://www.uspreventiveservicestaskforce.org/Page/Document/UpdateSummaryFinal/autism-spectrum-disorder-in-young-children-screening</a>. Accessed on February 9, 2018.
- 8. U.S. Preventive Services Task Force (USPSTF): Autism Spectrum Disorder in young children: Screening (2016). JAMA. 2016; 315(7):691-696.
- 9. U.S. Preventive Services Task Force (USPSTF): Screening for Speech and Language Delay in Preschool Children (2006). Available at: <a href="http://www.ahrq.gov/professionals/clinicians-providers/guidelines-recommendations/guide/section3.html">http://www.ahrq.gov/professionals/clinicians-providers/guidelines-recommendations/guide/section3.html</a>. Accessed on February 9, 2018.
- 10. Volkmar F, Siegle M, Woodbury-Smith M, et al.; American Academy of Child and Adolescent Psychiatry (AACAP) Committee on Quality Issues (CQI). Practice parameter for the assessment and treatment of children and adolescents with autism spectrum disorder. J Am Acad Child Adolesc Psychiatry, 2014; 53(2):237-257.
- 11. Yeargin-Allsopp M, Rice C, Karapurkar T, et al. Prevalence of autism in a US metropolitan area. JAMA. 2003; 289(1):49-55.

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

## Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

- 12. Zwaigenbaum L, Bauman ML, Fein D, et al. Early Screening of Autism Spectrum Disorder: Recommendations for Practice and Research. Pediatrics. 2015; 136(Suppl 1):S41-S59.
- 13. Zwaigenbaum L, Bauman ML, Stone WL, et al Early Identification of Autism Spectrum Disorder: Recommendations for Practice and Research. Pediatrics. 2015; 136(Suppl 1):S10-40.

#### Index

Asperger's Syndrome

Autism

Autism Spectrum Disorder

Fragile X

Pervasive Developmental Disorder (PDD)

#### History

Status	Date	Action
	09/20/2018	Updated Coding section with 10/01/2018 ICD-10-CM changes; added Z13.40-Z13.49.
Reviewed	02/27/2018	Medical Policy & Technology Assessment Committee (MPTAC) review.
Reviewed	02/23/2018	Behavioral Health Subcommittee review. The document header wording updated from "Current Effective Date" to "Publish Date." Updated References section. Updated Coding section to include codes 96101, 96102 96103.
Revised	08/03/2017	MPTAC review.
Revised	07/21/2017	Behavioral Health Subcommittee review. Revised document title. Updated formatting in Clinical Indications section. Revised MN criteria to align more closely with DSM-5. Updated References section.
Reviewed	08/04/2016	MPTAC review.
Reviewed	07/29/2016	Behavioral Health Subcommittee review. Updated formatting in Clinical Indications section. Removed ICD-9 codes from Coding section.
Reviewed	08/06/2015	MPTAC review.
Reviewed	07/31/2015	Behavioral Health Subcommittee review. Updated References section.
	02/02/2015	Clarified position statement. Replaced the term "mental retardation" with "intellectual developmental disorder."
	01/01/2015	Updated Coding section with 01/01/2015 CPT descriptor changes for 96110.
Reviewed	08/14/2014	MPTAC review.
Reviewed	08/08/2014	Behavioral Health Subcommittee review. Replaced the term "assessment" in document title and clinical indications section with the term "testing". Revised
	01/01/2014	Discussion and References sections.
	01/01/2014	Updated Coding section with 01/01/2014 CPT changes; removed 92506 deleted 12/31/2013.
Revised	08/08/2013	MPTAC review.
Revised	07/26/2013	Behavioral Health Subcommittee review. Revised title and clinical indications sections to replace "Pervasive Developmental Disorders" with "Autism

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

## Screening and Assessment for Autism Spectrum Disorders and Rett Syndrome

		Spectrum Disorders". Revised Description, Discussion, References, and Index
		sections.
	07/01/2013	Updated Coding section with 07/01/2013 CPT changes.
Reviewed	08/09/2012	MPTAC review.
Revised	08/03/2012	Behavioral Health Subcommittee review. Revised title to delete "Screening"
		and "Tools". Clarified role of other services in Clinical Indications section.
		Deleted quantitative plasma amino acid assays to detect phenylketonuria from
		MN section. Deleted radiological NMN services that are referred to in other
		company documents. Revised Discussion, References, and Index sections.
		Updated Coding section to remove CPT 84030 (no longer addressed).
	01/01/2012	Updated Coding section with 01/01/2012 CPT and HCPCS changes.
Reviewed	08/18/2011	MPTAC review.
Reviewed	08/12/2011	Behavioral Health Subcommittee review.
Revised	05/19/2011	MPTAC review. Moved content related to chromosomal microarray testing to
		new policy GENE.00021 Comparative genomic hybridization (CGH)
		microarray testing for developmental delay, autism spectrum disorder and
		mental retardation. Updated Coding section; removed 88384, 88385, 88386,
D : 1	11/10/2010	\$3870.
Revised	11/18/2010	MPTAC review. Added chromosomal microarray testing to not medically
		necessary section. Updated Rationale and Reference sections. Updated Coding
Reviewed	08/19/2010	section; removed 92569 deleted 12/31/2009. MPTAC review. Revised document title. Coding updated.
Reviewed	08/27/2009	MPTAC review. MPTAC review.
Reviewed	07/25/2008	MPTAC review. Deleted "and treatment" from medically necessary statement
Reviewed	07/23/2008	regarding services appropriate to screen for PDDs. Updated Discussion and
		Reference section.
Reviewed	11/29/2007	MPTAC review. Added Definitions section.
Reviewed	07/01/2007	Updated Coding section with 07/01/2007 HCPCS changes.
Reviewed	12/07/2006	MPTAC review.
New	12/01/2005	MPTAC initial guideline development.
		Sustained and sustained

This Clinical UM Guideline is intended to provide assistance in interpreting Healthy Blue's standard Medicaid benefit plan. When evaluating insurance coverage for the provision of medical care, federal, state and/or contractual requirements must be referenced, since these may limit or differ from the standard benefit plan. In the event of a conflict, the federal, state and/or contractual requirements for the applicable benefit plan coverage will govern. Healthy Blue reserves the right to modify its Policies and Guidelines as necessary and in accordance with legal and contractual requirements. This Clinical UM Guideline is provided for informational purposes. It does not constitute medical advice. Healthy Blue may also use tools and criteria developed by third parties, to assist us in administering health benefits. Healthy Blue's Policies and Guidelines are intended to be used in accordance with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.