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The Brazilian CARS: a standardized screening tool for autism

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Autism is not a disease. There is no blood test, neuroimaging, or electroencephalography (EEG) test for making or confirming this diagnosis. Autism is a behaviorally-defined syndrome that denotes atypical development of the immature brain and becomes manifest in late infancy, in toddlers, or young preschoolers. To the experienced clinician, classic autism is an obvious diagnosis. But experience and expertise are not measurable and therefore do not suffice when it comes to enrolling children in research studies or convincing a school district to provide expensive specialized educational services, especially to a child who has not yet celebrated his (or her) third birthday.

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The International Classification of Diseases (ICD-10)[1] and the American Diagnostic and Statistical Manual of Mental Disorders (DSM-IV)[2] were designed to be parallel, with minor differences between them. They specify that pervasive developmental disorders (PDD – autism for short in this paper) affect three behavioral domains: 1) sociability and empathy; 2) communicative language and imagination (pretend play); and 3) cognitive and behavioral flexibility. Symptoms must be apparent by age 3 years, be significantly handicapping and not explained by some other diagnosis, although there are no exclusionary criteria for a diagnosis of PDD/autism. The DSM provides descriptors of four behaviors in each of the three domains and specifies that for a diagnosis of autistic disorder (classic or Kanner's autism) no less than six of the descriptors must be endorsed, at least two in sociability, and at least one of each in language and play and in rigidity and perseveration. Fewer items, one of which must be sociability, or a different distribution of items yield a diagnosis of Asperger disorder if language developed at the usual age and the intelligence quotient (IQ) is at least 70, or PDD-not otherwise specified (PDD-NOS) for the remainder. We do not discuss here disintegrative disorder, for children who regress after fully normal early language and development, and Rett syndrome, a specific genetic cause of autism.

The diagnosis of autistic disorder is easy and uncontroversial when it occurs in an apparently healthy toddler, because of the salience of lack of social relatedness, impoverished play, repetitive apparently purposeless behaviors (stereotypies), lack of expressive language or else stilted, verbose, echolalic utterances, temper tantrums and aggressive behaviors elicited by intrusions into the child's activities, not to mention narrow food choices and sleep problems. It becomes much more difficult and controversial if autism is associated with attention deficit disorder with hyperactivity, severe cognitive deficiency, or high intelligence.

Behavioral diagnosis in childhood is rarely a yes/no dichotomy because criteria change with development which is discontinuous and variable in its rate. Behavioral diagnoses are perforce dimensional. Because of the lack of a sharp demarcation between the edges of normality and pathology, diagnostic reliability increases with increasing deviation from the population mean. Diagnosis therefore rests on agreed-upon rather than absolute criteria, and reliability mandates clinical instruments with strong psychometric properties.

Clinicians and researchers differ in their goals. Clinical diagnosis is forced to address all comers. It is concerned with the cause of all the presenting problems and with what to do about them. Because services are limited, clinical diagnosis is often shaded in borderline cases to ensure that children are not deprived of appropriate services by their diagnostic label. In contrast, researchers seek samples with strongly defined

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diagnoses and ages in order to maximize cross-site reliability, eliminate outliers, and reduce overlaps with control groups. The most widely used and best documented instrument since its publication in 1980 is Schopler's Childhood Autism Rating Scale (CARS).[3,4] It is considered the strongest rating scale for behaviors associated with autism. It has been translated in a number of languages because autism is a worldwide disorder.

The differing goals of clinicians and researchers affect the choice of diagnostic instruments and their uses. Screening instruments like the CARS are designed for efficiency in the clinic and for identification of potential research subjects. Screening requires clinical confirmation or, for research especially, corroboration with more comprehensive, demanding, and time-consuming diagnostic instruments.

The past quarter century has seen the development of a number of standardized questionnaires to parents and observations of children's behaviors and activities. The well-standardized Autism Diagnostic Interview-Revised (ADI-R),[5] which provides a historical perspective on autistic behaviors, and the Autism Diagnostic Observation Schedule-Generic (ADOS-G),[6] behavioral data using modules appropriate for different ages and developmental levels, are *de rigueur* for much current research. Their drawbacks are that these are very time-consuming, require expensive and extensive training, and are unrealistic and inappropriate for clinical and many research applications.

Some readers may ask why a Brazilian rather than a Portuguese CARS? No doubt for the same reason that American and British English, virtually identical in written form, have in three centuries drifted quite far apart in their vernaculars. The same no doubt occurred to Portuguese, as Pedro Álvares Cabral landed on the Brazilian coast already in 1500, preceding colonization of the United States by English speakers by some 80 years. Thus Brazil's longer Europeanized and deeply multicultural history starts earlier than that of the United States, which no doubt enriched its oral language to an even greater degree. It is not only appropriate but required, therefore, that behavioral instruments be adapted to the dialect and also the mores and vernacular of the country in which it will be used.

Pereira et al.[7] provide an exemplary translation and standardization of the CARS for Brazil: two independent translators who compared notes to reach a unified version, followed by its translation back into English by a third. To establish its sensitivity and specificity, the Brazilian CARS was then given to a standardizing population of 60 Brazilian children referred to a university diagnostic clinic in the city of Porto Alegre, southern Brazil, ages 3-17 years, 73% of them boys as is typical of autism. The investigators did not include in their standardizing population typically developing children of the Porto Alegre region or other Brazilian cities (after all, Brazil is a huge country and there must be significant differences between north and south, coastal and inland cities and regions), nor children with nonautistic developmental disorders. The cautionary words of the authors suggest such studies may be forthcoming.

Strengths of the CARS are that it requires relatively little training, has been in use for over 30 years, was standardized in large (n = 1,500) populations in the United States, and is used in multiple countries. Rather than DSM/ICD system diagnoses based on the 12 descriptors of behaviors in three behavioral domains, the CARS provides a four-point severity scale (absent, mild, moderate, severe deficit) for each of 14 well-described behaviors, plus a single overall diagnostic severity score. The overall sum CARS score ranges from a potential of zero (no autistic features) to 60 (all severe features endorsed).

Further work indicated that scores of 36-60 indicate severe autism (i.e., autistic disorder), 30-35 moderate autism, < 30 not autism. One can criticize the CARS because it does not separate Asperger syndrome from PDD-NOS and because there are children with autism features among those with scores in the 20s who do not make the cut for moderate autism. Some clinicians would argue that they are on the spectrum, in the gray zone between "normal" and "mildly affected." These borderline cases overlap with cognitive impairment, obsessive-compulsive disorder, semantic-pragmatic language disorder without autism, and other co-morbid phenotypes.

High convergence of the Brazilian CARS with the Autistic Traits Assessment Scale[8] and with clinical diagnosis indicates that it is identifying children with autism appropriately. High internal consistency of the 14 behaviors that target social reciprocity, communication, and restricted and repetitive behaviors supports autism as a single construct. We were surprised not to see any examination of the effect of age or IQ on the psychometric properties of the Brazilian CARS. Lord[9] showed, in her follow-up study of 2-year-olds referred for a possible diagnosis of autism, a tendency for the CARS to overdiagnose autism in cognitively handicapped children at 2 years but less so at 3. Another weakness of the CARS is its unreliable discrimination of young children with autism from mental-age matched children with other disorders, especially limited language.[9]

The Pereira et al. paper[7] is a model of how to adapt a well-accepted test to a new culture, which we hope readers from other countries will emulate. We also hope that the international community of scholars will accept forthcoming clinical and experimental studies of autism from Brazil which use the Brazilian CARS as part of the diagnostic process. It is important, however, that not only obvious cases be diagnosed but also more controversial cases, especially mildly affected children for whom appropriate intensive early intervention may spell the difference between ability to adapt and function in the real world and an empty expensive life of

dependence. The CARS will not fulfill all the needs of such children, but it is clearly a realistic and important first step in this direction for Brazil.

References

1. World Health Organization. The ICD-10 classification of mental and behavioural disorders. Clinical descriptions and diagnostic guidelines. 10 ed. Geneva: World Health Organization; 1992.
2. American Psychiatric Association. Diagnostic and statistical manual of mental disorders. Fourth edition, text revision: DSM IV-TR. 4th ed. Washington, DC: American Psychiatric Association; 2000.
3. Schopler E, Reichler RJ, DeVellis RF, Daly K. Toward objective classification of childhood autism: Childhood Autism Rating Scale (CARS). J Autism Dev Disord. 1980;10:91-103.
4. Schopler E, Reichler RJ, Renner BR. The Childhood Autism Rating Scale (CARS) for diagnostic screening and classification in autism. New York: Irvington; 1986.
5. Lord C, Rutter M, Le Couteur A. Autism Diagnostic Interview-Revised: a revised version of a diagnostic interview for caregivers of individuals with possible pervasive developmental disorders. J Autism Dev Disord. 1994;24: 659-85.
6. Lord C, Risi S, Lambrecht L, Cook EH Jr, Leventhal BL, DiLavore PC, et al. The autism observation schedule-generic: a standard measure of social and communication deficits associated with spectrum of autism. J Autism Dev Disord. 2000; 30:205-23.
7. Pereira A, Riesgo RS, Wagner MB. Childhood autism: translation and validation of the Childhood Autism Rating Scale for use in Brazil. J Pediatr (Rio J). 2008;84:487-494.
8. Ballabriga MCJ, Escudé RMC, Llaberia ED. Escala d'avaluació dels trests autistes (A.T.A.): validez y fiabilidad de una escala para el examen de las conductas autistas. Rev Psiquiatr Infanto-Juvenil. 1994;4:254-63.
9. Lord C. Follow-up of two-year-olds referred for possible autism. J Child Psychol Psychiatry. 1995;36:1365-82.

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Asthma in early life: is the hygiene hypothesis correct?

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Since the original studies in the late 1980's there have been literally hundreds of birth cohort studies that have purported to document the role of respiratory infection and genetic susceptibility in the development of early life asthma, including the paper in this issue looking at wheezing in southern Brazil and explain the results based on the hygiene hypothesis.[1]

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As originally formulated by Strachan, the hygiene hypothesis suggested that younger siblings had less hay fever than their older siblings because they had more frequent infections and hence less allergy.[2] Von Mutius, and others have popularized this hypothesis, as the explanation for the asthma epidemic.[3] In my view, there is substantial contradictory data to suggest that both the hygiene hypothesis itself, and its ability to explain the asthma epidemic are not correct.

- Point 1: Although there is an ecologic relationship between the decline in all infections and the rise of T helper 2 (Th2) diseases, the hygiene hypothesis does not explain why T helper 1 (Th1) autoimmune diseases have increased over the same time frame as the Th2 diseases.

- Point 2: As pointed out by Platts-Mills, and others, the decline in infections diseases is a result of antibiotics, better housing, better water, and this decline antedated by many years the epidemic of both Th1 and Th2 autoimmune diseases.[4]

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