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Special Issue: Autism

What's in a Name???

GOOD - authentic, honest, just, kind, pleasant, skillful, valid

NEIGHBOR - friend, near

ALLIANCE - affiliation, association, marriage, relationship

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Commentaries

Authorship

In October, Peter Rintels, MD, a hematologist, oncologist in private practice wrote to express concern on behalf of himself and his colleagues about a case reported in this journal (*Medicine & Health/RI* 2004;87 (8)). His view is published in this journal.

He and his colleagues had not been notified about this report in advance. A teaching exercise summarizing a morbidity and mortality conference on hemophagocytic lymphohistiocytosis had failed to mention the work done by him and his associates in diagnosing and caring for the patient. The authors, both chief residents, had not cared for the patient themselves, and had not acknowledged the efforts of the doctors who had been responsible for the patient. Nor had they acknowledged the efforts of the numerous other doctors who had worked to heal the patient. To add insult to injury, some of the statements about management were deemed incorrect.

The real problem was the lack of acknowledgement to the community doctors. There was a perception that the full time academic staff would not have been so slighted but the private practitioners could be ignored. The case report wasn't reviewed by the experts who cared for the patient, so that manuscript errors were not detected, the end result being a somewhat inaccurate report and several wounded psyches. "We are very pleased to see physicians in training preparing difficult cases for publication so the lessons learned can be shared with the wider community… We hope that in the future a more thoughtful editorial policy will not trivialize the efforts of those of us for whom this is more than mere intellectual exercise."

I spoke with Dr. Rintels. I am a strong believer in eliminating town-gown tension. I have recruited editors for issues of this journal from both sides of the divide, although definitely more from the academic side for a number of reasons. This journal represents the Rhode Island Medical Society and I try to maintain that perspective. Dr. Rintel's letter raised some interesting issues, however, and we ultimately agreed that the real problem was the one of acknowledgement, not authorship.

Let me deal with authorship first. As an author and co-author of several case reports, I can only confirm the general experience that writing the manuscript is often easier than dealing with the other doctors about who "owns" the case. The more interesting patients have often been cared for by multiple services, with several fellows, attendings and teams of housestaff. Each may have a particular point of view, reminiscent of the five blind men describing an elephant. Who gets to write up a case? Who gets to be a co-author? How many authors are too many? Can you submit a report on a single case with ten authors? 15? Should the intern who actually did all the work get the credit even though he had no idea what was going on? What about the resident or the fellow? What if the case was "solved" by an attending who spent little time or effort but was able to put the pieces together? It is not so uncommon to find an interesting case reported by different specialties in different journals by non-overlapping teams of doctors, none of whom knew the other was writing up the case. In recent years journals have established criteria for authorship, but these are commonly overlooked, especially with case reports, when authorship can be claimed based simply on caring for the patient in some capacity.

In the case at hand, however, we are dealing with a teaching exercise based on a teaching conference on a single case, to make a number of teaching points. This is not a case report. It is a teaching module. Nothing new was described, simply a review of arcane but useful information. Dr. Rintels and I agreed that he and his colleagues should not have been authors. Should they have been offered the chance? I think that if the goal had been to write a case report, the answer is yes, assuming they contributed to the write-up of the case (NOT for caring for the patient. That was their job.) But for a teaching exercise, I think not and Dr. Rintels agrees.

But should they have been contacted? Certainly. It seems like common courtesy to tell a doctor that his case was going to be the subject of a report, just as the doctors should have been told that the patient would be the subject of a teaching conference. Yet it can be embarrassing for one doctor to tell another that the case is being written up but that the doctor is not invited to participate. Publications are, after all, one measure of academic accomplishment, so that every publication has merit, regardless of how little merit the publication deserves. And this oversight may be particularly galling to community physicians who, not long before, were themselves the academic kings of their environment.

Another interesting issue is the responsibility of the journal to the patient. In a state as small as Rhode Island, where people with rare disorders may be readily identified, should the journal have confirmed a written informed consent from the patient? Several years ago the *New England Journal of Medicine* published a case report from the University of Michigan. Shortly thereafter the subject of the case report, who had been anonymous, wrote an irate, signed letter to the *Journal* about this breach of his anonymity. Although the anonymity seemed to have been punctured by the patient himself, rather than the case report, the *Journal* decided that right was on his side and now requires written consent for case descriptions. I do not agree with this approach and would only require written consent if the description of the disorder made the patient's identification quite apparent.

The solution that I've come up with is to ask the authors of these teaching exercises to contact the doctors who cared for the patient to let them know of the manuscript, and to have them review it. In addition the principal players should be acknowledged in the manuscript. Will this solve the problem? No. Some doctors will think of themselves as having been more involved than others will have deemed them, and it will often be difficult to determine who actually took care of a patient who spent three weeks in the ICU, where 30 different doctors may have signed the chart, often illegibly.

As editor of this journal I am as upset as I am pleased, on the one hand that I failed to consider this problem before it arose, while on the other, that people are reading the journal and care enough to express their concerns. I hope the latter will continue.

-Joseph H. Friedman,MD

Old Age, Stricken in Years

An 1886 issue of the *Lancet* carried a brief article by Dr. Jonathan Hutchinson describing a patient of his, a six year-old boy. The child's birth had been unremarkable but, by age one, the infant's growth slowed perceptibly; he became bald and wizened. By age six he weighed but 20 pounds and had all of the outer appearances of an aged man. His skin, for example, was thickened, excessively wrinkled and had lost the elasticity associated with youth. It seemed as though the child had skipped adolescence and proceeded, without interruption, into an accelerated senescence.

Yet another case of premature aging in a child surfaced about 20 years later; and gradually, physicians came to recognize a specific disorder of childhood, fortunately rare, characterized by rapid aging. Based upon a Greek word for aging, they called it progeria.

It was a bizarre disorder, with virtually all of the customary stigmata of aging, including cataracts of the eyes, premature deafness, baldness, and even accelerated sclerosis of the coronary and cerebral arteries. But the morbid fascination which this rare hereditary disease provided for medicine went deeper. Physicians wondered whether the secret mechanisms of normal aging might somehow be encapsulated, and then exaggerated, in these afflicted children. Perhaps, in their plight [progeric children rarely live beyond age 16], these youngsters might harbor the mysterious mechanisms, though amplified, underlying the aging process affecting the remainder of human society. And further, if the disease process in progeric children, whatever it is, can be slowed or even neutralized, such knowledge might be applicable to the bulk of humanity as it undergoes what is called "normal" aging. Some geneticists have even dared to wonder whether the "normal" aging is not, in reality, a reversible disease afflicting all.

Certainly aging is not uniformly applied to all who survive childhood. Some humans age more rapidly than others [males, for example, more than females]. And in the absence of major trauma, both genetic inheritance as well as life-style seem to determine jointly the span of life allotted to each human. Life-style, of course, embraces a multitude of factors, including prudent diet, physical exercise, avoidance of ambient pollution, avoidance of major emotional stress and social strife and a conscious intent to live a purposeful life.

As medical science delved more deeply into the countless metabolic pathways and interactions which serve the functioning vertebrate body, inevitably a search has been initiated to explain the phenomenon of aging. Why, for example, do large mammals, such as elephants, live beyond 70, while little mammals, such as mice, live at most three years? Physiologists, pondering the varied life expectancies of a wide assortment of creatures, have sought some rational explanation for this wide diversity. They noted that the ratio between the creature's volume and its skin area is proportional to its average life-span. Still other observers observed that the number of heart beats during a mouse's life is about equal to the number of heart beats during an elephant's span of year. Small creatures, it seems, live fast and die young while the hulking animals creep slowly through a much longer life. In the words of the ecologist Brian Enquist, "You can spend it all at once or slowly dribble it over a long time."

Scientists first distinguished between life-span, the inexorable biological limits of life, under the best of circumstances; and life-expectancy which represents the actual duration of life. The latter is reality and the former, the ideal limits of life.

The critical biological factors modulating the process of aging, however, have remained elusive. Environmental circumstances clearly determine why Norwegian women live, on average, to age 83 while Ethiopian woman survive, on average, to age 35 years. The thought that aging is merely the residue of a cumulative wear and tear process, in some settings more stressful and life-shortening than in others, seemed unsatisfactory.

How then, it was asked, do stress, poor diet and exposure to certain chemicals accelerate aging? Recent findings indicate a variety of separate avenues of inquiry, each quite promising, including the role of chemicals involved in the metabolism of connective tissue, the role of free radicals, cumulative errors in the production of body proteins, mutational changes in the body's DNA, mistakes in the body's immune response such that it may attack its own tissues as though they were foreign invaders; and, of course, the inheritance of certain genes which may create additional time limits on the life span.

Progeria is a rare event, arising about once in every eight million births. And other than the victims and their parents, the disease has not prompted much public health concern. Yet science has extensively investigated this disorder seeking for a specific gene variant which can be held responsible for the "fast-forward" velocity of aging in these children.

In 2003 a team of scientists at the National Institutes of Health, aided by investigators from a number of universities including Brown, identified the genetic mutation responsible for childhood progeria, the most dramatic form of premature aging. [This abnormal gene, called LMNA, plays an important role in coding for critical cell membrane proteins.] Francis Collins, the Director of the national Human Genome Research Institute, declared: "This genetic discovery represents the first piece in solving the tragic puzzle of progeria. The implications of our work may extend far beyond progeria – to each and every human being. What we learn about the molecular basis of this model of premature aging may provide us with a better understanding of what occurs in the body as we all grow older."

In recent years, often employing the procedure called "positional cloning," geneticists have identified the genes responsible for cystic fibrosis, Huntington's disease and a number of other hereditary maladies. Inevitably, ethicists will also explore the societal implications of extending active adult life for a decade or two. Seneca, who knew no genetics, said, however, that old age is an incurable disease.

-Stanley M. Aronson, MD

Introduction

Joseph J. Hallett, MD

Autism has become popular with the media and public. We have heard that autism is increasing at an alarming rate. A study by the US Department of Education found a 100% increase in autistic children in some schools districts. In 2003 Rhode Island Kids Count reported that autism has increased dramatically in Rhode Island. There are individuals who travel the lecture circuit discussing their experience with autism. This rush towards autism has placed **primary care physicians (PCPs),** parents, schools and community governments in a situation of increasing demands for diagnosis, treatment and education for a disorder that they don't understand well and have heard conflicting claims about.

This issue aspires to provide its reader and in particular the PCP with a better understanding of autistic disorder and autistic spectrum disorder. A perspective on what at times appears as an elusive diagnosis is provided by a brief history in autism, its terminology and its epidemiology. Drs. Gargus and Yatchmink discuss the early identification of at-risk children. As with all disorders of brain development, screening is directed at identifying infants or children who fail to express specific behaviors (e.g., language, speech, social) at a particular age. The early diagnosis of children with autism allows early intervention and family support. Identification of at-risk children is not complex, validated screening tools are available and PCPs can combined them with their current period screening of motor, language and adaptive development.

Autistic behavior is complex and can appear to lack a central organizing neurocogntive process. Dr. Sheinkopf discusses several hypotheses of brain dysfunction in autism. Central to the formulation of these hypotheses is emerging data on the social brain and the neural processes that facilitate social interaction.

The PCP must provide care of children with autism. As with many chronic disorders, autism will utilize all a PCP's skills. Dr Burke and his colleagues discuss the role of PCPs and the problems that confront them. The need for a PCP in childhood to treat illness, advocate and comfort extends into adults, when new social and health problems arise. The PCP must care for both. It can be said that individuals with autism are best served by the medical community when they have a PCP home. This issue aspires to enable home-building.

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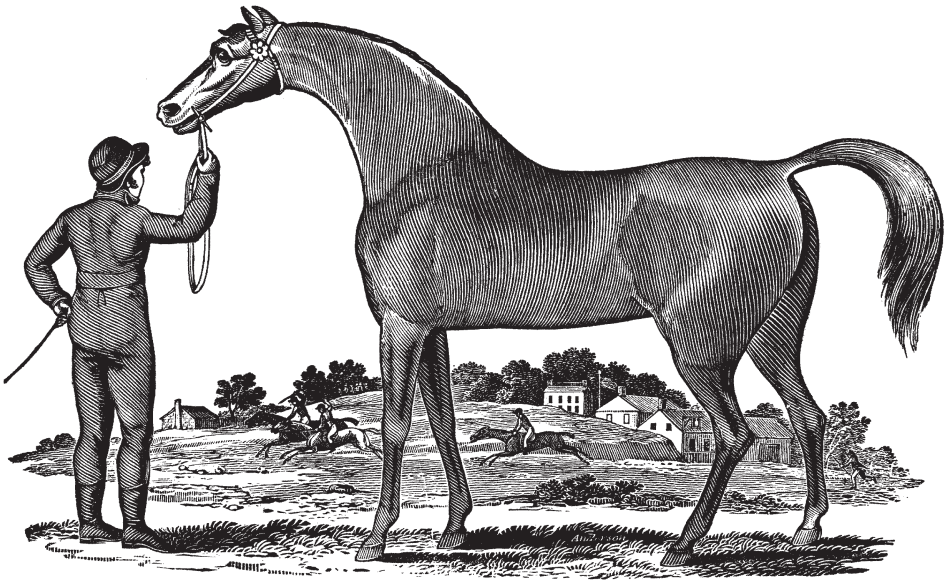

Making Sense of Autism

Joseph J. Hallett, MD, and Viren D'Sa, MD

Autistic disorder (AD) is a disorder of brain development. Its neuropathogenesis and etiology are thought to be heterogeneous and are know in only a small fraction of cases. Genes have a complex indirect and, to date, undefined role in AD. This chronic neurologic disorder leaves affected individuals dependent on their families for life.

Terminology

Autism, autistic disorder and autistic spectrum disorder need definitions. They are best understood from a historical perspective.

In 1943 Leo Kanner, a child psychiatrist, described 11 children, most from prosperous and well-educated parents (several were physicians) with "autistic disturbances of affected contact."[1] He borrowed the word autistic from the schizophrenia literature, where it denoted the self-centered thinking, detached behaviors and resulting isolation observed in schizophrenia. Following his report, the term infantile autism emerged.

Kanner's report received little attention in part because the behavioral phenotype was already subsumed under the diagnosis of childhood schizophrenia. However, in the 1950s the psychodynamic theorists resurrected his work and hypothesized that children became autistic because they were not given appropriate emotional nurturing by their "refrigerator" mothers.[2,3] This approach to autism was the antithesis of Kanner's speculation that infantile autism had a biologic cause. Infantile autism was formalized as a disorder in 1980 when it was included in the **Diagnostic Statistic Manual, third edition (DMS III).**[4] It was categorized as a **pervasive developmental disorder (PPD),** a group of behaviorally defined syndromes that share the characteristic of severe and pervasive disruption of development in "..social interaction skills, communication skills or the presence of stereotyped behavior, interests and activities."[4] The other PDDs are Rett's disorder, childhood disintegrative disorder, Asperger's disorder and **PDD not otherwise specified (PDD NOS**). PDD NOS was included to allow clinicians to indicate that a child had a behavioral phenotype that was similar to one of the defined PDDs but did not meet the full criteria for that disorder. In the next revision of the DSM III infantile autism was changed to **autistic disorder (AD**), the term used in subsequent editions, including the DSM IV.

With the explosion of research in the past 25 years, the concept has emerged that the behavioral characteristics observed in children with AD are not unique to AD but are distributed in lesser forms and in different combinations throughout a population; AD behaviors represent a portion of this distribution. In the 1990s the descriptive term, **autistic spectrum disorder (ASD)**, was coined in the experimental literature. The term migrated into clinical literature where it is often used as a diagnostic term even though a common clinical definition has not been agreed upon.

> **"AD and the other PDDs are behaviorally defined syndromes that medicine and the sciences are struggling to define with quantitative, objective diagnostic criteria ..."**

Pervasive Developmental Disorders

AD and the other PDDs are behaviorally defined syndromes that medicine and the sciences are struggling to define with quantitative, objective diagnostic criteria and to associate the phenotype with a biological marker. At present neither is available although the **Autistic Diagnostic Observation Schedule (ADOS)** offers a meaningful advancement. Diagnostic criteria are based on a qualitative determination of the severity of behavioral impairments, and demonstrating that a child has the DSM IV prescribed number of impairments. This qualitative determination is subject to the confounding of observer bias and marked rater-to-rater variance. This confounding variance has been a recurring source of considerable disagreement among physicians, a disagreement that no doubt has perplexed the PCP who is caring for the child and supporting the family. The ADOS offers an observational instrument to reduce the influence of these confounding variables.

Autistic Disorder

AD is a behaviorally defined syndrome characterized by severe impairments in social interaction, the social components of language and inflexible and restricted behaviors or interests. A diagnosis is made based on a behavior phenotype defined in DSM IV.[5] Since the diagnosis is based on behaviors, the AD phenotype can overlap with other disorders. For example, a large percent of children with tuberous sclerosis have the AD phenotype. Similarly, approximately 1% of children with AD have tuberous sclerosis. This overlap of phenotype is also seen with congenital brain malformations and untreated phenylketonuria. However, most cases of AD are idiopathic.

Autistic Spectrum Disorder

The basic behavioral traits that comprise autism have not changed since Kanner reported them. However, the interpretation of these traits has changed and so has the type of child receiving an autism diagnosis. The

broadest interpretation is represented by the term **autistic spectrum disorder (ASD)**. It assumes that autism is not a discreet disorder but should be viewed as a spectrum of conditions or behaviors unified by impairments in social interactions, communication and restricted behaviors or interests. Since ASD is defined by behaviors and not by pathogenesis or causative agent, a spectrum can include the Kanner-type AD in which there is severe and profound impairments as well as mental retardation, to milder cases such a PDD NOS, to atypical cases such as high functioning AD (AD with IQ > 70), to cases with no impairment of language development and only mild impairments in social interaction and inflexibility such as in Asperger disorder, to children whose impairments in the past were considered below diagnostic threshold for PDD. The dilemma present by this approach is how to distinguish between ASD behavior and behavior that is eccentric, idiosyncratic or odd which are in general considered within the range of normal.

Support for interpreting autism as ASD comes from studies that find AD behavioral traits, albeit with varying severity, distributed through families of children with AD and in the general population.[6, 7, 8] Further support comes from functional brain imaging studies (fMRI) that found similar activation patterns in adults with high functioning autism and Asperger syndrome.[9] On the other hand, the term spectrum is most often used in the context of disorders that are clinically distinct and share a common etiology while varying in the severity of symptoms. The validity of grouping behaviorally defined phenotypes of potentially different etiologies has been questioned. AD and the disorders subsumed under ASD are postulated to have heterogeneous etiologies which open the possibility that the autistic behavioral phenotype may represent a final common pathway for a variety of disorders. Determining whether an autistic phenotype represents the final common expression of distinct disorders or represents a spectrum of disorders with shared neuropathological parameters is a question awaiting clarifying studies.

Although the utility of ASD is debated, it has become a popular clinical term, used to define a behavioral phenotype without reference to pathogenesis or etiology. An understanding of this is important because the use of ASD must be accompanied by a vigorous diagnostic evaluation for treatable disorders.

Epidemiology

The epidemiology of AD has not changed substantially from the findings reported in the 1980s and early 1990s. It is most often diagnosed in males (male to female ratio of 4 to 1). Cases do not segregate by race, level of education, socioeconomic status or geography. Behavioral signs typically appear before the age of three.[10] Delays in language (either absence or slow progression) are the most common presenting complaint.[11] A small portion have regression after typical language development. Sixty to seventy-five percent of the children with AD have mental retardation.[12,13] Cognitive deficits may be more severe in girls than boys although approximately 40% of boys will have severe to profound mental retardation. Organic, behavioral and emotional comorbidities are common. (Table 1)

AD has implications for the entire family. Mild language, social or psychiatric problems can be present in parents.[14, 15] Siblings have a greater, albeit small, risk of AD, language disorder, learning disabilities, social problems and psychiatric disorders.

Prevalence

A rise in the prevalence of children with an autistic diagnosis has been established although a specific prevalence rate has not.[12,16,17] Population studies in the 1980s and early 1990s found a prevalence ranging from 2 to 10 per 10,000 children.[13, 17] After 1994 prevalence increased annually with recent estimates reaching as high as 67 per 10,000 for ASD and 4 per 10,000 for AD.[12] Since 1994 Rhode Island has mirrored this trend in its special education population.[18] In 2003, 605 students with an autistic diagnosis were educated in Rhode Island schools. Understanding the factors behind this rise in prevalence is critical to our understanding of AD and ASD.

Population-based incidence studies, using contemporary diagnostic criteria, are believed to more accurately reflect potential changes in the occurrence of disorders across time periods. One such

Table 1. Comorbidities in autistic disorder

Mental retardation (60-75%)
Epilepsy (5% in children, 30% in adults)
Phenylketonuria if untreated (5%)
Tuberous sclerosis (<1%)
Neurofibromatosis
Congenital malformations
Cerebral palsy
Down syndrome
Hearing impairments
Vision impairments
Learning disabilities
Sleep disorder
Self-injurious behaviors
Aggressive, angry or combative behaviors
Oppositional behavior
Hyperkinetic behavior
Depression
Anxiety
Obsessive compulsive disorder
Tics

study in **Olmsted County Minnesota (OC)** found increases in the incidence of AD from 0.55 per 10,000 children in 1980-1983 to 4.49 per 10,000 children in 1995-1997.[17] A rising incidence among young children was responsible for most of the increased. During the same period the incidence among children older than 10 years of age remained stable. Several putative environmental agents, such as mercury and vaccines, have been intensely studied, but to date no environmental agent has been identified to explain the rise in incidence. Genetic and genomic studies have also failed to explain the rise.

Prevalence studies assessing time trends are subject to inaccuracies that result from changes in diagnostic criteria, inability to validate diagnoses and increased awareness of the disorder across time, all of which have occurred in autism.[17, 19, 20] The broadening of the diagnostic criteria can be seen in the revisions of the DSM since infantile autism was first included in 1980. The use of ASD has broadened the criteria still further. Public and physician awareness has grown substantially. Validating the AD or ASD is short coming in many prevalence studies. A second study of **Minnesota children (MS**) illustrates the problems. This study found a greater increase in prevalence, to 52 per 10,000, than the OC study which measured incidence.[21] This difference can be explained in part by the method of ascertaining subjects and validation differences between the two studies. In the OC study, investigators reviewed medical and school records to determine the diagnosis of each subject using DSM criteria. The MS study used student data reported to the department of education. Such data do not allow diagnostic validation.

The possibility that school data may be biased towards categorizing students as autistic is suggested by the timing of the initial rise in prevalence (between 1991 and 1994 in most studies). Coincident with the initial rise was the inclusion of autism in the list of disabilities eligible for federally mandated special education services in 1991. If this biased diagnosis, studies ascertaining cases using school data would be expected to identify a rise in prevalence. Support for this possibility would be evidence of diagnosis swapping. Croen[20] found evidence of diagnosis swapping in California, a state that has reported a dramatic rise in autism. A period of rising prevalence in AD saw a corresponding drop in the prevalence of mental retardation without autism.

The reasons for the disturbing rise in the prevalence of AD and ASD do not suggest a meaningful rise in new cases but a rise due to diagnosis swapping, an increased identification of cases and a broadening of diagnostic criteria to include cases that were considered below the diagnostic threshold in the past. However, the possibility that a portion of the rising prevalence results from a true increased incidence in autism has not been excluded.

Table 2. Examples of disorders associated with autistic disorder.

in utero exposure to rubella, CMV, valproate, thalidomide

neonatal or early infant insults (e.g., hypoxia, infection, trauma, hypothyroidism)

brain malformations

chromosomal abnormalities (5%)

Gene mutations (1-2%)
- 15q11-13 (Prader Willi/Angelman region) most frequent
- fragile X mutation
- tuberous sclerosis
- phenylketonuria (if untreated)

Etiology

AD is categorized as either idiopathic or secondary when the cause is known. Approximately 90% of the cases are idiopathic. A cause of the AS can be identified in approximately 10%. (Table 2) Chromosomal abnormalities account for 5% of the secondary AD. Duplication in the Prader Willi/ Angelman region (15q11-13) being the most frequent gene mutation. Untreated phenylketonuria was responsible a considerable portion of secondary AD in the past but is now rare. Other causes of AD include prenatal agents such as infection and hypoxia. Although several environmental agents have been implicated as causative agents (e.g., mercury), studies have not provided supporting evidence.

Current conceptualization of the role of genes in AD is that of imparting susceptibility but not causing AD. Genetic heterogeneity is the operating hypothesis in studies with estimates of the number of involved genes ranging from 10-15. Four candidate susceptibility genes are currently under study.[22] (Table 3) Linkage studies have identified chromosomes 2, 7, 17, 22 and X, but confirming studies have given inconsistent results. Numerous candidate genes have been identified including most neurotransmitter receptors and proteins for essential brain development (e.g., neuroligin, reelin), but no candidate gene has been consistently found in children with AD. This lack of clarity is not inconsistent with overlapping gene effects. Genes can be epistatic (several genes influencing a behavior) or pleiotrophic (one gene influencing several behaviors).

Inheritance

The inheritance of idiopathic AD is complex and non-mendelian. No single inheritance pattern has been recognized. Inheritance is thought to be multifactoral involving the interaction

Table 3. Candidate autism susceptibility genes

	chromosome locus	
AUTS1	7q11	(William syndrome region)
AUTS2	3q25	
AUTS3	13q14	
AUTS4	15q11	(Prader-Willi/Angelman syndrome region)

of genes and epigenetic factors. The evidence for a genetic component in the neuropathogenesis of AD comes from twin and family studies. Twin studies have demonstrated a higher concordance in monozygotic twins than dizygotic. [23] Furthermore, some family pedigrees show an increased recurrence risk within families with one child with AD.[24]

The recurrence risk to siblings of a child with secondary AD is the risk of the disorder causing AD (e.g., tuberous sclerosis). Determining the empiric risk of idiopathic AD depends on the incidence of AD which is debated (see Prevalence). Barbaresi reported an incidence of 4.5/10,000.[17] Using this incidence, the empiric risk is less than 0.1%. Some have recommended using prevalence which would raise the risk to 0.5-0.7%. The risk in ASD is debated but is presumed to be greater than AD. The risk increases considerably in families with one child with AD (4%) and still more when there are two or more children with AD (35%).[24] Because of the association between AD and language, social and psychiatric problems, families with one child with AD are given an additional risk of 4-6% for one of these problems.

Neuropathogenesis

Kanner was the first to speculate that AD had a biological cause. He supported his speculation by observing that several children had large heads. His observation of macrocephaly has been confirmed by anthropomorphic and neuroimaging studies. Macrocephaly is present in 20% of the children with AD, appears late in the first year and resolves by five years old in most children. The high incidence of seizures and mental retardation provides further evidence for a neuropathogenesis.

Studies to elucidate abnormal brain regions in AD have produced inconsistent results while implicating numerous cerebral, cerebellar and brainstem regions. Failure to replicate neuroanatomical studies, in part because of the unavailability of postmortem brain tissue, has hampered an understanding of how brain development is disrupted. The etiologic heterogeneity of AD may also explain inconsistencies between studies. **Functional magnetic resonance imaging (fMRI)** offered investigators one method of circumventing the paucity of brain tissue although it is still limited by etiologic heterogeneity. Despite this, it has provided a means of correlating brain dysfunction with brain structures.

Since a hallmark of AD is impaired social components of language and cognition (discussed by Dr. Sheinkopf in this issue), the "social brain" region has received particular attention. [25] Social cognition can be thought of as the ability to recognize, manipulate and behaviorally respond to social information whether in the form of language, another's behaviors or expressions. An important pathway mediating social cognition involves the amygdala, superior temporal sulcus and fusiform gyrus of the orbitofrontal cortex. fMRI studies have demonstrated the abnormal activation of these structures in high functioning autistic individuals.

Of particular interest is the serotonin system: 30% of children with AD have elevated platelet serotonin. Whether this contributes to the neuropathogenesis of AD or is an incidental finding will require further study.

Studies of the autistic brain have produced heterogeneous collections of findings most likely the result of its heterogeneous etiologies. The picture that appears to be emerging is disruption of early brain development with subsequent abnormalities in neuronal morphology and number, synaptic abnormalities and dysfunctional pathways as a consequence.

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Early Identification and Assessment of Young Children with Autism

Regina A. Gargus, MD, and Yvette Yatchmink, MD, PhD

When should autism be diagnosed?

A growing body of evidence supports the benefit of targeted early intervention for children with autism spectrum disorders.[1] Experts maintain that children should be identified sometime between 18 and 24 months of age.[2] Most children with autism spectrum disorders show signs of social-emotional behavioral impairment in infancy. [3] The majority are non-dysmorphic in appearance, and have normal growth patterns. Therefore, the primary care provider should recognize the red flags of atypical social-emotional development. [Table 1, Table 2].[3,4] If any of these flags are identified, the child should be referred for more comprehensive assessment. Early identification supports families,[5] allows primary care providers to offer insights for management, and helps parents and other care providers to understand the child's strengths, learning preferences, challenges and behavioral problems [e.g.: delayed toilet training, temper tantrums and sleep disorders].

"In the course of a year, a primary care provider will see between 3 and 7 children with an autism spectrum disorder for every 1000 children in their practice."

Where should assessment for autism take place?

The American Academy of Pediatrics endorsed the "Practice Parameter: Screening And Diagnosis Of Autism" developed by the American Academy of Neurology and The Child Neurology Society.[4] Routine surveillance, defined as a flexible, continuous process whereby professionals perform skilled observations of children during the provision of health care,[6] helps identify children at risk for any type of atypical development. Surveillance should include: [1] addressing parental concerns, [2] obtaining a developmental history, [3] direct observation of the child, and [4] use of surveillance tools. Good surveillance tools have a sensitivity of 70-80%; therefore, a normal evaluation result, while encouraging, still requires reevaluation at the next well-child visit. When a problem is readily observable, the child should be immediately referred for an assessment to a specialist at an autism diagnostic center based on the primary care physician's clinical judgment, because most screening tools have a70-85% specificity. Children

TABLE 1: Typical and atypical infant social emotional and language development

Expected typical Infant Social-Emotional And Language Developmental Milestones [4]	Red Flags Of Atypical Infant Social-Emotional And Language Development [3]
4mo: shows interest in watching people's faces, smiles back, initiates smiles **6 mo**: relates to parents with real joy, smiles with parents during play **9mo**.: back and forth smiles/sounds/gestures, give & take activities **12 mo**: orients to name, uses gestures to get needs met, plays peek-a-boo/patty cake, repeats actions clapped for **15mo**: checks parent's facial expression after unexpected stimulus and then reacts accordingly, uses sounds pointing and showing gestures to draw attention to objects of interest, begins to show empathy [becomes concerned when others cry. **18mo**: simple pretend play [feeding doll with play bottle] and attracts parents by looking up at them or gesturing during play **24mo**: more complex pretend play [wooden peg represents bottle to feed baby doll] or engages in 2 step pretend play [feeds doll then puts doll to sleep] without prompting from adult; enjoys being next to other children, shows interest in playing, offers another child a toy **36 mo**: imagines self as a different character; talks for doll or action figure; plays with other children; shows and tells another child about a favorite toy; talks about feelings [hungry, sad, sleepy] past and future.	If your Baby shows any of these signs, ask your pediatrician or family practitioner for an immediate evaluation: • No big smiles or other warm, joyful expressions by six months or thereafter. • No back and forth sharing of sounds, smiles, or other facial expressions by nine months or thereafter. • No babbling by 12 months. • No back and forth gestures, such as pointing, showing, reaching, or waving by 12 months • No words by 16 months • No meaningful phrases [without imitating or repeating] by 24 months. • ANY loss of speech or babbling or social skills at ANY age.

TABLE 2: Social-communication skills in young children and in autism

DEFINITIONS	NORMAL DEVELOPMENTAL PRESENTATION
Joint Attention[26] is defined as the ability to coordinate one's own attention between an object and another person. Joint attention is a pre-linguistic triad between the child, caretaker, and object that advance from non-verbal following of caretaker gaze to object; proto-imperative period of using gaze between object and caretaker to meet needs; following finger pointing to proto-declarative use of pointing finger[29] with gesture, words, or gaze to bring caretakers attention to object of child's desire/need. *Parents of children with autism find they are often self directed, self engaged and lack showing and sharing behaviors.*	8m: gaze monitoring 10-12m: follow point 12-14m: isolates own finger for pointing 14-16m: show / share by pointing
Social Orienting [27,28] is the ability to respond to social cues or stimuli such as turning in response to own name; engagement of others in social setting. P*arents of children with autism may question possible hearing loss in their child..*	8-10m: responds to name 10-12m: looks at faces of others; shares objects
Symbolic Play[29] is pretend play and correlates to receptive and expressive language, requires communication, and is a social skill. *Many children with moderate to severe autism remain at the sensory motor phase, mouthing objects, using stereotypic-non-intended ways to interface with toy[lining up behavior], banging, throwing, spinning or twirling toys*	8-10m: [sensory motor] bangs /throws block or toy 12-14 m: uses toys for intended purpose –builds tower with blocks 16-18m: [simple] use objects representatively [talk into toy telephone] 18-20m: [complex] pretend with any generic object [shoe may be put to ear to represent telephone]
Theory of mind[30] Child aware that others have thoughts and feelings different from own, can learn from others. *Children with autism often are rigid in their behaviors and routines, unable to comfortably accept change.*	30-36 m children have the ability to take another person's perspective

referred to diagnostic centers have a higher incidence of developmental problems, even when the evaluation does not result in a diagnosis of autistic disorder, Asperger's disorder or Pervasive Developmental Disorder, not otherwise specified.[7] Surveillance tools should evaluate all major domains of child development (social-emotional, gross and fine motor, communication and language, general cognitive and knowledge, and self help-behavior-adaptive functioning). A combination of surveillance tools frequently gives the most comprehensive assessment of a child's level of functioning, and serial reassessment at each pediatric visit provides the best opportunity for early identification and timely referral for further assessment.

When developmental deficits are noted, the child should be promptly evaluated with formal audiologic assessment, lead testing, and screened with tools specifically for the identification of disorders on the autism spectrum. A positive autism screen should result in prompt referral of the child to clinicians experienced in the evaluation and treatment of ASD.[8] In addition, the child should be given a referral to Early Intervention or the local public school department for educational, language and social skills interventions. Table 3 provides a summary of instruments available for general developmental surveillance, and autism specific screening. Surveillance and screening tools must provide ease of use for the parent and the healthcare provider, have minimal cost for materials and administration, and have an established age appropriate standardization and validation for the instrument; informal tools lack the sensitivity and specificity for early detection.[9]

There is no gold standard for developmental surveillance or autism screening, but several instruments have reasonable psychometric properties. The **Age and Stage Questionnaire (ASQ)** can be completed by parents and has reasonable sensitivity and specificity. The **Parents Evaluation of Developmental States (PEDS)** (Table 3) is easy to administer, with strong psychometrics. The **Modified Checklist for autism in Toddlers (M-CHAT)** has become increasingly popular as an autism screen. The Society for Developmental and Behavioral Pediatrics recently endorsed the **Primary Care Screener (PDDST II PCS –stage 1).**

Once a child is referred to a center, personnel may use instruments such as the **Autism Diagnostic Interview revised (ADI-R)** in conjunction with the **Autism in Diagnostic Observation Schedule (ADOS),**[10-13] The ADOS allows direct observation of behavior by trained personnel as they offer structured and unstructured activities to elicit social and communication behaviors that are frequently difficult for individuals with autism spectrum disorders. Modules were designed to evaluate nonverbal children as well as verbal toddlers to adults. Although the original validation sample consisted of only 381 subjects, this instrument is now used for diagnostic confirmation and has become the benchmark measure in almost every research study involving autistic children.

Why is early identification by primary care providers important?

Approximately 25% of children in a primary care practice present with developmental concerns.[4] The American Academy of Pediatrics recommends that the primary care physician provide a medical home with comprehensive health supervision, which includes developmental and behavioral surveillance at every pediatric visit. [14] Parents should be encouraged to discuss

TABLE 3: Some Surveillance Tools and Autism Screening Tools for Primary Care Offices

Test	Description	Author/source
SURVEILLANCE TOOLS FOR PCP OFFICE		
ASQ : Ages and Stages Questionnaire **ASQ-SE**: Ages and Stages Questionnaire Social-emotional [6m-60m] English/Spanish/ others	15 min-Parent completed Child monitoring system 30 items: Problem Solving, Communication, Gross Motor, Fine Motor, Personal –social skills ASQ : Sensitivity: 70-90%; Specificity: 79-91%; Interrater reliability 0.56-0.87 Cost/Child: $4.60-9.20 material + admin ASQ-SE: Sensitivity: 75-89%; Specificity: 79-90%; Interrater reliability 0.94	Carnahan,S,PhD. Katz, R. 2002 Paul,H. Brookes www.brookspublishing.com
IDI Infant Development Inventory [3-18 months] English/Spanish	10 min Parent completed. Infant monitoring system with 60 yes-no descriptions for five domains. Sensitivity 75% Specificity 70% Cost/child: $3.80 mat+admin	Ireton,H. PhD 1996 www.childdevrev.com
CDI Child Development Inventory [15m-72m]	30-50min: Parent completed 320 questions assesses development, symptoms and behavior problems of young children in relation to age norms and identifies the child's strengths and areas of delay. Profile of 8 developmental scales evaluating domains of social, self help, gross motor, fine motor, expressive language, language comprehension, letters, numbers, and general development.. Sensitivity 100% specificity 94-96%	Ireton, H & Glascoe, FP [32] 1995 www.agsnet.com
PEDS Parents Evaluation of Developmental Status [Birth –8 years] English/Spanish	2 min: Parents completed 10 questions response form looking at cognitive, language, motor, behavior, social-emotional and self help skills related to typical development, does not identify specific impairment. Sensitivity: 74-79%. Specificity:70-80% Cost/child: $1.19 mat+admin	Ellsworth & Vanderer Press 2001 www.pedstest.com electronic format www.forepath.org
Brigance Screens [0-90 months] Infant –Toddler:0-23m Early Preschool:24-30m Preschool:30- 54m K-1 Screen: 55-90m	10 min Parent completed. Nine separate forms for each 12 month age range. speech, language, social-emotional, motor, general knowledge and readiness. Sensitivity: 70-87%; Specificity:70-82%; interrater reliability 0.9. Cost/child: $11.68 mat+admin	Brigance A.N. 1985 [new version 2005] www.curriculumassociates.com
PSC Pediatric Symptom Checklist [4-18 years] English, Spanish	35 short statements of emotional problem behaviors including externalizing [conduct, attention, etc.] and internalizing [anxiety, adjustment, depression, etc.]. Sensitivity 95% middle income, 88% lower income. Specificity 68% middle income/100% lower income.	Jellinek, M and Murphy, JM [33] 1999 http://psc.partners.org for free download of measures. www.pedstest.com for free copy of factor scoring
SCREENING TOOLS FOR PCP OFFICE		
M-CHAT Modified Checklist for Autism in Toddlers [24-36 months]	5 min. Report of 23 yes-no questions evaluating social communication behaviors. Sensitivity: 90%; Specificity: 99%; Cost/child $0.98	Robbins, D. etal [34] 2001 www.firstsigns.org/downloads/m-chat.pdf Free
CHAT Checklist for Autism in Toddlers [18 months]	Short questionnaire with 14 items focused on behaviors which when absent at 18 months put child at risk for a social-communication disorder. Sensitivity 65-85% Specificity 100%	Baron-Cohon [35] 1992 www.nas.org.uk
PDDST II PCS –stage 1 Primary Care Screener [12-18 months]	For PCP office to answer question of should there be a substantial concern about possible autism needing referral. Sensitivity 92%; Specificity 91%	Siegel, B [36] 2004 www.PsychCorp.com
CSBS DP Communication and Symbolic Behavior Scales Developmental Profile [6 to 24 months]	Measure of seven language predictors in young children for communicative competence: emotion and eye gaze, communication, gestures, sounds, words, understanding and object use. Sensitivity: 88.9-94.4% Specificity 88.9% Interrater reliability 0.92-0.97	Wetherby, A. & Prizant B. www.pbrookes.com
SCQ Social Communication Questionnaire [4y or older, with cognitive age> 2y]	10 min. Parent Completed. Previously known as Autism Screening Questionnaire. Instrument evaluates communication skills and social functioning contains 40 items derived from the ADI-R. Sensitivity: 96% Specificity: 80%	Berumet, SK. Rutter,Michael, etal [37] 1999 www.wpspublish.com

The sensitivity of a developmental tool is the probability that it will correctly identify children who exhibit developmental delays or disorders. The specificity of a developmental tool is the probability that it will correctly identify children who are developing normally.[31]

TABLE 4: **Dignostic Center Tools For Autism Assessment**		
PDDST II DCS –stage 2 Developmental Clinic Screener [birth –18months]	Designed for developmental pediatric practice to answer question: Should additional autism specific assessment be carried out on this child. Sensitivity: 73% Specificity 49%	Siegel, B [36] 2004 www.PsychCorp.com
PDDST II ACSC –stage 3 Autism Clinic Screener [birth-18 m]	For use in clinics where autism is regularly diagnosed, looks at severity of autism spectrum disorder. Sensitivity: 58% ; Specificity 60%	Siegel, B [36]2004 www.PsychCorp.com
ADI-R Autism Diagnostic Interview-Revised [18m-adults]	Standardised, semi-structured diagnostic interview for use with the parents or caregivers of people with suspected autism or Asperger's Disorder. The interview focuses upon three main areas (i) quality of reciprocal social interaction, (ii) communication and language, and (iii) repetitive, restricted and stereotyped patterns of behaviour	Lord,C. Rutter, LeCouter [38] 1994 www.wpspublish.com
ADOS Autism Diagnostic Observation Schedule [toddlers-adults]	30-45 min by Trained Personnel. A standardized protocol for direct observation of social and communicative behavior . It uses a variety of structured and unstructured activities to elicit a wide range of behaviors associated with autism spectrum disorders. There are four modules , each directed to a particular developmental age and language ability [nonverbal to verbally fluent]. ADOS does not distinguish Asperger Disorder from Autism Disorder.. Autism vs other: Mod 1-4 Sensitivity: 93-100%; Specificity:93-100% Autism +ASD vs Other: Mod 1-4: Sensitivity 90-97; Specificity:87-94% Autism vs ASD + Other: Mod 1-4: Sensitivity 87-100% Specificity: 68-79% . ASD vs Other: Mod 1-4 Sensitivity 80-94% Specificity 88-94% Interrater reliability 0.7-0.92	Lord,C. , RisiS etal [18]2000 www.wpspublish.com
CARS Childhood Autism Rating Scale [Age 24-73 mn]	15 items covering aspects of behavior that are abnormal in children with Autism Sensitivity: Autism vs. PDD/nonPDD: 64%; ASD vs. nonASD: 47%; Autism vs. PDD.NOS: 90%; CARS vs. Cl Dx: 86% Specificity: Autism vs. PDD/nonPDD: 92%; ASD vs. nonASD 94%; Autism vs. PDD. NOS: 100%; CARS vs. Cl Dx: 91%. Interrater reliability 0.63	Schopler,E., Reichler,RL., Renner,B www.agsnet.com
GARS Gilliam Autism Rating Scale [3 years-22 years]	5-10 min: Identify and diagnose autism and estimate severity. 42 test items grouped into 3 subtests: stereotyped behaviors, communication, social interaction, to describe specific, observable and measurable behaviors; and 4th subtest of 14 items on developmental disturbances for parents to contribute data about their child's first 3 years of life. Interrater reliability 0.8-0.9 Sensitivity 48% [39] Interpretation: Test generates an Autism Quotient [AQ], standard scales with mean 100 and standard deviation +/-15. 50% of patients with Autistic Disorder have an AQ between 90-110 91% of patients had an AQ >80 [range indicative of probable Autistic] An AQ>110 is range highly indicative of Autistic Disorder.	Gilliam,J. 1995 www.agsnet.com

The sensitivity of a developmental tool is the probability that it will correctly identify children who exhibit developmental delays or disorders. The specificity of a developmental tool is the probability that it will correctly identify children who are developing normally.[31]

TABLE 5: Resources for Professionals and Parents

- Autism Society of America www.autism-society.org
- First Signs [education about ASD] www.firstsigns.org
- National Alliance for Autism Research [NAAR] www.naar.org
- OASIS Online Asperger Syndrome Information and Support www.aspergeersyndrome.org

- Rhode Island Technical Assistance Project, Autism Center. Sue Constable. 401-222-4600 x2014
- The Autism Project of Rhode Island www.theautismproject.org
- The Autism Society of Rhode Island acastle2@netzero.net
- Families for Early Autism Treatment of Rhode Island [FEAT/RI] www.featri.org
- Asperger's Association of New England www.anne.org

developmental questions [15] during their well child visit. Parental concern about motor, language, cognitive, school or global development has been shown to be highly sensitive [79%] for the identification of disabilities. [16] However, many first- time parents lack a frame of reference to recognize developmental delays. They depend on their primary care provider to identify these delays.[17] Handouts that outline developmental expectations[18] can assist families in following the developmental progress of infants and toddlers. The primary care provider has at least 12 opportunities for developmental surveillance during the first three years of the child's life.

Final Thoughts

In the course of a year, a primary care provider will see between 3 and 7 children with an autism spectrum disorder for every 1000 children in their practice. Adopting effective routine developmental surveillance and specific autism screening for at-risk children increases access to early treatment and improves potential outcomes for children and their families.[19] The National Survey of Early Childhood Health noted that only 42% of parents of children in the 10-35 month age group recall being told that a developmental assessment was being conducted and 57% of parents recall that their child received a developmental assessment.[20,21] Long term child and societal costs are substantially reduced by providing ongoing developmental surveillance, screening, and referral.[22,23]

The American Academy of Pediatrics, the **Centers for Disease Control and Prevention (CDC),** and First Signs (discussed by Dr. Burke et al in this issue) have endorsed the **A.L.A.R.M**. campaign. [24] A.L.A.R.M. seeks to raise awareness of **A**utism prevalence, encourage providers to **L**isten to parents, **A**ct early by making surveillance and screening a routine part of practice, **R**efer for definitive diagnosis and intervention, and **M**onitor progress and ongoing access to resources. [Table 5]. Early referral to intensive [15 to 20 hours per week], structured and generalizable behavior management strategies to address communication and social skills, and incorporating curricula that promote development of Joint Attention skills improves the potential for young children. [25]

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Hot Topics in Autism: Cognitive Deficits, Cognitive Style, and Joint Attention Dysfunction

Stephen J. Sheinkopf, PhD

Autism is characterized by severe deficits in a range of social skills and behaviors. In many ways it is possible to think of autism as a social learning disability. However, other disorders of childhood, especially other developmental disabilities, also affect social development. In addition, there is a growing consensus that children with autism represent a heterogeneous group. Thus, one can view autism as a collection of phenotypes, perhaps varying along a continuum, perhaps having multiple etiologies with converging characteristics, or perhaps having divergent phenotypes from common etiologies. So it is a daunting task to identify a narrow range of characteristics that can help physicians screen for the disorder.

With these caveats in mind, several features of autism are unique to the disorder. They appear to be good indicators of autism in early life, and offer insight into both the severity and quality of the social deficits in autism. In this brief paper, I will focus mainly on one aspect of social functioning in autism; namely, deficits in joint attention. However, I will also briefly review other "hot topics" in autism research, including deficits in social cognition (including theory of mind), executive functions, and central coherence. Each of these concepts will be considered in relation to the joint attention deficits seen in autism.

Joint Attention

The Normal Case

Joint attention is a family of social abilities ("social pragmatic abilities") that help individuals regulate, respond to, and engage in social interactions with others. Of particular interest to the study of autism are **joint attention behaviors (JA)** that regulate social interactions between a child, an interactive partner, and other aspects of the environment. These types of joint attention events are termed triadic social interactions.[1]

Throughout the first two years of life, infants develop increasingly complex nonverbal, vocal, and (sooner for some than others) verbal skills that can be deployed to meet the demands of social interactions. There are a number of different ways to view the repertoire of social pragmatic behaviors in this early developmental period. Infants may initiate social interactions, or respond to social bids of others. Social-communication episodes may also vary with respect to *communicative function*. (Figure 1) Children may use nonverbal behaviors to request an object out of reach, or they may request help with an object. The means to this social end may vary, with some acts involving pointing, others involving eye contact, others simply a reach, and some combining a number of behaviors. Whatever the form, however, these behaviors serve as requests, for they have some instrumental value and function to elicit aid from a social partner.

Other behaviors coordinate attention between a child, a social partner and the environment in order to share social information. These acts do not have the same instrumental function as requests and instead serve a social-affiliative or *commenting function*. Indeed, these acts are sometimes termed "protodeclarative."[2] In this paper I call these acts "joint attention" acts. More specifically, infant-initiated joint attention acts are termed **initiating joint attention (IJA).** The responsive form of this class of social pragmatic behavior is termed responding to **joint attention (RJA***)* and reflects the ability of infants to orient their gaze or focus of attention to that of a social partner, as when an infants looks to where an adult is pointing.

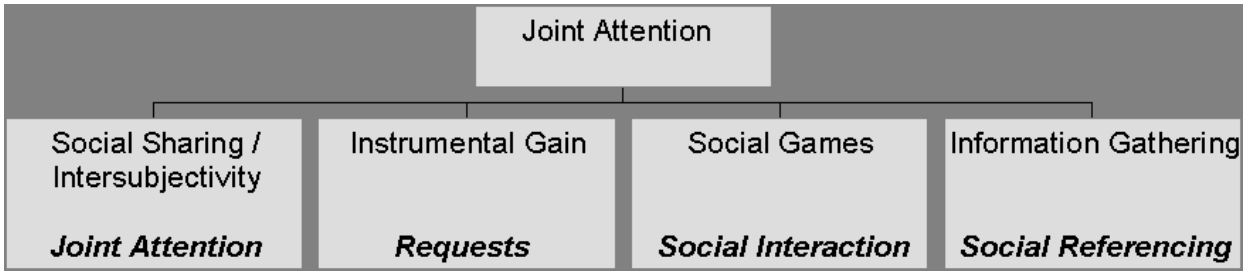
Joint Attention in Autism.

Autism is characterized by severe deficits in both IJA and RJA.[3-5] Children with autism show deficits in the frequency and complexity of joint attention behaviors, as well as differences in the qualitative patterns of strengths and weaknesses in joint attention as related to other social pragmatic behaviors. Autism is characterized by generally poor social and communication skills. But there is a particular weakness in IJA and RJA skills. Thus, children with autism show severe deficits in the frequency with which they **initiate joint attention bids (IJA)** as well in their ability to monitor and respond to **joint attention bids of others (RJA).** These deficits contrast with a relative sparing of requesting skills.

This is not say that the children with autism show completely normal and age-appropriate abilities to request and to make their needs known. Indeed, children with autism may show deficits in the complexity of requests (e.g., failure to use pointing or poor coordination of eye contact, vocalizations, and other communicative means when making requests). Instead, the pattern of social pragmatic skills seen in autism, particularly in young children with autism, reflects a greater propensity to regulate the behavior of others for instrumental gain and a diminished tendency and/or ability to coordinate attention with others for social affiliative functions.[6]

Such patterns of social and communicative abilities can be seen through behavior observation and are evident in parents' descriptions of their children. How do we observe these behaviors in the laboratory or clinic? In the laboratory, we stage a semi-structured play session where a clinician presents a series of toys and games designed to elicit requesting and joint attention behaviors.[7] The clinician will present a toy that creates a spectacle, like a wind-up toy. While this toy is active (e.g., moving about, hopping, making noises, etc), a typically developing infant may point, alternate gaze with

Figure 1: Functional uses of joint attention behaviors



the examiner, smile, and/or vocalize, all in the service of coordinating attention between themselves, the adult, and the toy. When the infant touches the object, s/he may even hold it up to *show* it to the adult. These are all bids of joint attention (specifically, ITA). When the toy stops, the infant may use eye contact to indicate that the adult should activate the toy again. More complex requests may involve points, eye contact, or even the child giving the toy to the adult. At other times, the infant may point to an object out of reach to request a new toy. Typically developing infants display both IJA and requests with some frequency, even in interactions with an unknown adult in a laboratory. In contrast, IJA acts are rare for young children with autism, whereas rudimentary requesting behaviors would be much more likely.

Joint attention deficits are important to our understanding of autism and to the identification and diagnosis of the disorder. The diagnostic criteria for autism (i.e., ICD-10 and DSM-IV) reveal a number of symptoms that are more or less reflective of deficits in joint attention. These deficits are among the earliest appearing symptoms[8] and are strongly represented in the scoring algorithms of screening and diagnostic instruments.[9,11] Joint attention deficits are related to the overall severity of symptoms in autism[12] and to cognitive and language development in this population.[13] Clinical experience indicates that this is a particularly difficult set of abilities to remediate through intervention, although recent efforts have targeted joint attention skills in interventions for young children with autism.[14]

Social Cognitive Deficits in Autism

A large body of literature on **Theory of Mind (ToM)** probes the ability of individuals to reason about the intentions and beliefs of others.[15] A seminal paper by Baron-Cohen and colleagues[16] reported that individuals with autism were unable to reason about the false beliefs of protagonists in a series of vignettes. This deficit has been widely replicated in subsequent research. One influential theory argues that ToM abilities are rooted in a discrete information-processing ability that allows children to represent the thoughts of others.[17]

Theoretical links between social cognition and joint attention include propositions of JA as a precursor to ToM abilities[18,19] as well propositions that JA is an early form or behavioral indicator of ToM.[20,21] There is a general assumption that for children to engage in acts of JA they must understand that the other has a unique point of view. JA and ToM have been argued to recruit a common cognitive module that lets an individual represent the thoughts and beliefs of others,[22] with JA involving a more basic and rudimentary form of understanding than later developing ToM abilities.[23] Such a view postulates that autistic deficits in pretend play, an additional marker of autism in early childhood, also share this requirement for representational thought.

Despite these theoretical links, there is little empirical evidence for the link between JA and ToM, in spite of some longitudinal links that have been reported.[24]

Executive Function Deficits.

Executive functions (EF) are a broad class of cognitive abilities involved in the regulation of thought and action. This class of higher cognitive abilities supports such functions as strategic planning, impulse control, working memory, organization of mean-end behaviors, and flexibility in thought and action. The frontal lobes are heavily involved in these processes.[25]

Deficits in EF are well replicated in children with autistic disorder.[26-28] However, research indicates that autism is related to a specific pattern of deficits in executive skills; i.e., deficits in planning efficiency and perseverative responses that indicate difficulties in shifting response set.[26,27] These patterns have been found to differentiate the executive performance of autistic individuals from those with ADHD and other neurodevelopmental disorders. Children with ADHD are most likely to show deficits in response inhibition, whereas autistic individuals tend not to show evidence of such dysfunction on EF batteries.[26,29]

An important issue for the EF deficits in this population is their developmental course. Ozonoff and colleagues have reported that deficits in planning efficiency and set shifting, purportedly related to prefrontal functioning, were of greater magnitude for older than for younger individuals.[27] This is consistent with the view that frontal lobes (and EF) are late to mature and suggests that floor effects may mask the appearance of such deficits at younger ages.

While EF deficits are most robustly seen in older and higher functioning individuals, at least one report documents increased perseverative responses on an object search task in preschool age children with autism.[30] Thus, it may be that early EF deficits may be seen with appropriately sensitive tasks. An interesting trend from the

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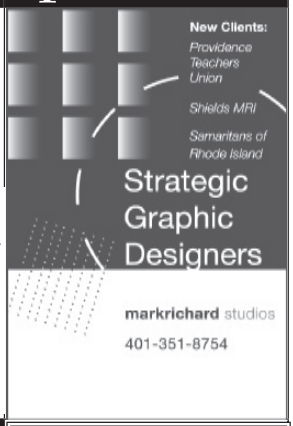

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studies of EF skills in younger children with autism is that performance differences on EF tasks appear to be related to early JA abilities.[30-32] In short, there may be a syndrome-specific set of EF deficits in autism, at least some aspects of which may be related to JA.

Central Coherence

Frith and Happé[33] proposed the concept of central coherence as an addition to the ToM approach in understanding the cognitive style of individuals with autism. Central coherence (the tendency to integrate information to form a whole, coherent meaning.) reflects a cognitive style that allows one to "see the forest for the trees." The central coherence theory is intended to help explain aspects of autistic functioning that are not well explained by ToM deficits; namely, the tendency to focus on local features of the environment.[34] In part, this theory was also intended to help explain why individuals with autism perform better on certain cognitive tasks; e.g., the embedded figures task, where an individual is required to identify a figure (e.g., a triangle) embedded in a meaningful picture. A local processing style is thought to favor this task and is predicted to result in faster responses (i.e., shorter latencies to find the target).

Empirical research on this concept is limited. Nonetheless, studies have found evidence for a preference for local versus global processing style in young children with autism (i.e., under age 5 years), and examined this construct in relation to JA skills.[34,35] There is also some evidence that relatives of children with autism (i.e., parents and siblings) may show a tendency towards this local processing style.[36] One obvious question is whether this processing style is related to, or perhaps a different level of explanation of, the executive dysfunctions described above. Very little data can be brought to bear on this question. One study has indicated that EF deficits and weak central coherence may be independent features of autism,[37] but this hypothesis remains unresolved.

A second question is whether weak central coherence is related to the social deficits seen in autism, and to deficits in JA. It has been suggested that weak central coherence limits an individual's ability to integrate aspects of the social world into a meaningful whole.[34,35] Some preliminary evidence suggests a relationship between central coherence and ToM abilities in typically developing and autistic children.[34] In addition, one report on JA and central coherence in children with autism and a comparison sample with developmental delay did not find evidence for a link between joint attention and central coherence.[35] Although it was not clear that the JA measure was an appropriate test of the JA deficit in autism, these results raise the hypothesis that weak central coherence may be independent of other social and cognitive deficits seen in autism.

Summary

This review has touched on selected hot topic issues in autism research. There are other exciting developments in the field, including advances in neuroimaging and genetics. Such advances notwithstanding, an understanding of the social and cognitive features of autism reviewed here has great importance. For example, research in our laboratory is focused on identifying factors that may underlie the JA deficit in autism. A better understanding of these factors would improve predictions about the presentation of the disorder in early infancy, as well as better target interventions on pivotal skills and behaviors.[38]

In addition to implications for research, concepts such as JA, ToM, EF, and central coherence can help health care providers develop a fuller picture of both the strengths and impairments that characterize autism spectrum disorders. This can help providers better understand autism not as a collection of isolated symptoms, but as a description of a population of children with syndrome-specific strengths and weakness.

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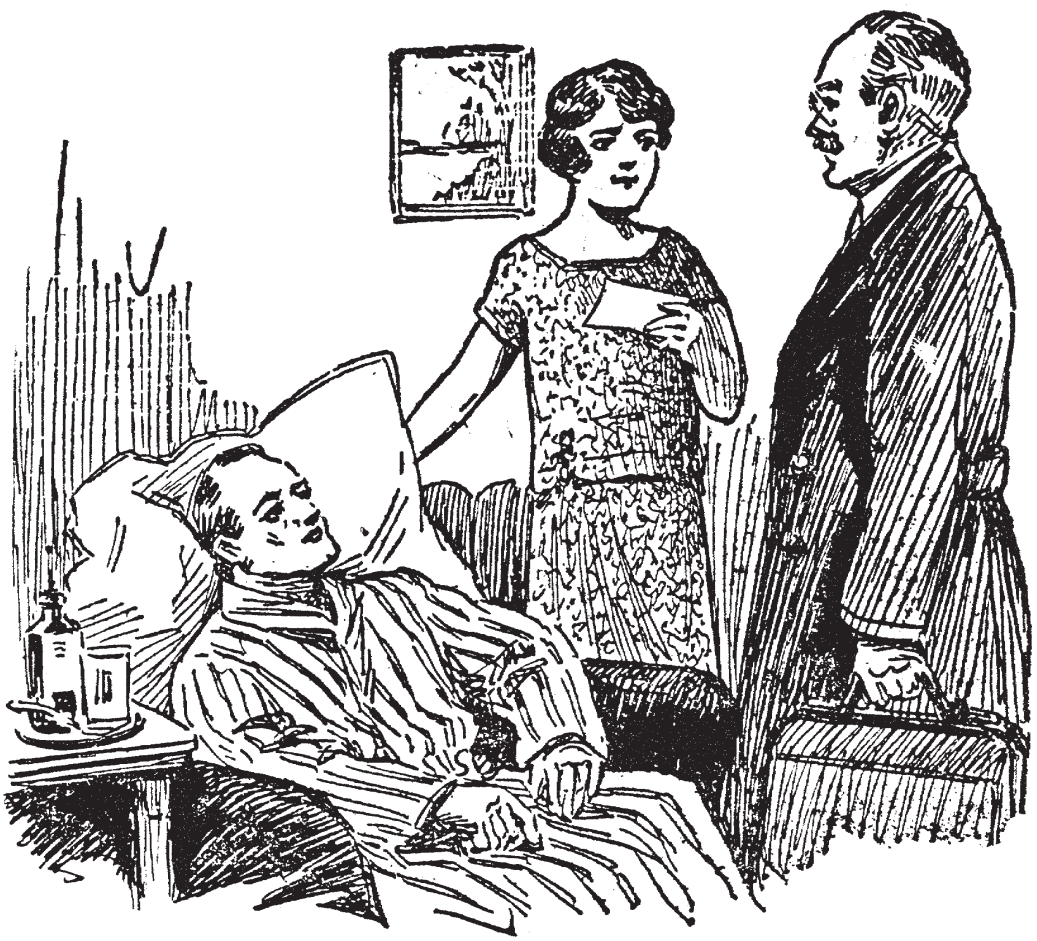

The Primary Care of Children with Autism

Robert T. Burke, MD, Ann-Marie Cardosi, RN, BSN,
Ashley Price, MD, FAAFP, and Alanna Teatom-Burke

Over the last decade, the subject of autism has been magnified in the minds of the general public, as well as in the consciousness of child development, education, medicine, and public health professionals. Autism, once thought of as rare, is now recognized as occurring much more commonly than believed and affecting not only individual children and families but the health care, early intervention and educational systems as well. In the not too distant past, autism was thought to effect approximately 4 or 5 children in 10,000.[1] More recent studies have reported incidence of 1 to 2 per 1,000 with some surveys reporting even higher incidences.[2,3,4] In Rhode Island, the number of children between ages 3 and 21 years receiving special education services who reportedly have one of the autistic conditions increased from 30 to 605 between 1993 and 2002.[5] The number of children in Rhode Island with one of the autistic conditions is estimated as being well over one thousand. Virtually every pediatric practice is likely to have at least one child with an autistic condition.

Autism and **autistic spectrum disorders (ASD)** represent a heterogeneous group of disorders with marked variability in the presenting characteristics of qualitative differences in reciprocal social interaction and communication and with restrictive behaviors that become apparent in early childhood. These children present the pediatric care provider with challenges in screening, diagnosis, treatment and management. Both residents in training and practicing pediatricians report autistic children among the more challenging groups of patients for whom they provide care. Nevertheless, most providers remain willing to provide care and to improve the level of care that they provide.[6, 7]

Over the last decade, The American Academy of Pediatrics, through its Medical Home Project, has promoted the role of pediatricians and family physicians in the care of children with and without special health care needs.[8] Among the provisions of the Medical Home are a range of clinical and supportive services.[9] These are the provision of primary health care including surveillance and screening, care management, referral and coordination of care, education and guidance for the child and family, advocacy and support and the transition of health care as the child matures.[10] The American Academy of Pediatrics has published an extensive technical report on the pediatrician's role in the diagnosis and care of children with any of the autistic spectrum disorders.[11] The Academy has also joined with the United States Department of Health and Human Services and other organizations to promote the A.L.A.R.M. Project to improve professional understanding of autism and to encourage screening and early referral for diagnosis and intervention.[12,13]

Increasing awareness of autism and autistic spectrum disorders is perhaps the most important initial step. The first "A" in A.L.A.R.M. indicates that "Autism is prevalent." Autism, Autistic Spectrum Disorders and other developmental disorders occur with greater frequency than previously believed: autistic disorders occur in more than one child in five hundred while other developmental or behavioral disorders may occur in as many as one child in six.[2]

"L" denotes " listening to parents". Parents of children diagnosed with autism at an age older than 3 years frequently have reported concerns about their child's development to a health provider by the time the child is eighteen months old; but parents report lengthy delays between reporting their concerns and referral. Parents' concerns may sometimes not point to a specific developmental disorder, but, more often than not, do indicate the need for more formal screening. Surveillance by asking questions related to child development should be part of the routine health maintenance examination for all children. This will improve the early identification of children with developmental problems when the parents do not report any specific concerns.

The second "A". stands for Acting Early. Some general surveillance questions can be red flags for identifying children at risk of developmental disorders. (Table 1)

A more formal screening process for Autistic Spectrum Disorder can be carried out during routine well-child examinations or selectively for children thought to be at risk based on answers to surveillance. (See the discussion by Drs. Gargus and Yatchmink this issue). Because there is an increased risk of an approximately 10% occurrence among the siblings of children with autism, the health care provider should monitor the social, communication, adaptive and behavioral development of the siblings of autistic children, not only for signs of autism but other cognitive or developmental disorders as well.

The next recommended step in A.L.A.R.M. is "R" for referral of children who are at risk for any developmental disorder, including Autistic Spectrum Disorders, to an Early Intervention program and to a developmental specialist for a diagnostic evaluation. The primary care provider can move this process along by obtaining an audiologic examination of hearing and a speech and language evaluation. Referral should be made as soon as a developmental risk is identified. This should be done even prior to the formal diagnosis of developmental disorder. Referral should also be made to a developmental specialist for a definitive diagnostic evaluation. This will ensure that the child will be promptly evaluated and enrolled in therapeutic services while the family receives support services.

Becuase autism is a complex and

Table 1.
Red Flag Screening Questions for Autistic Spectrum Disorders

*No babbling by 12 months of age
*No pointing or other gestures by 12 months
*No single words by 16 months
*No two-word sentences by 24 months
*Any loss of language or social skill at any age.

Other questions and observations focus more directly on autistic spectrum disorders, including; "Is your child able to:

*communicate as well as other children his/her age?
*show good eye to eye contact?
*respond to his/her name?
*interact with people like other children his/her age?
*smile back at people reciprocally?
*wave bye-bye?
*point to objects to draw your attention to them?
*tell or show you what he/she wants or does he/she have to lead you by the hand to get things?
*bring you books or toys of interest to him/her simply to show you?
*play interactively with other children?
*play in a way that is typical of other children his/her age and gender?
*play with toys in a typical way?
*engage in pretend play if over 2 years of age?
*have the ability to calm him or herself in a relatively short time when upset or having a tantrum?
*get to sleep and remain sleeping all night?

"No" or negative answers indicate the need for further evaluation.

multifaceted condition, the definitive diagnosis and characterization of specific disabilities is best carried out by a team of experienced evaluators.[15] Referral to a pediatric developmental specialist or autism diagnostic unit should be made as soon as possible to clarify the diagnosis and to document the child's developmental and behavioral challenges. The diagnostic assessment should be based on formal diagnostic criteria such as those published in the DSM-IV or the ICD-9. The **Autism Diagnostic Interview - Revised (ADI-R)** and the **Autism Diagnostic Observational Scales (ADOS)** [16,17,18] are not only useful in establishing the diagnosis of Autistic Spectrum Disorder, but are invaluable in documenting the behavioral and developmental challenges that will need to be addressed in any behavioral or educational service plan.

Referrals to Early Intervention for children under age three and to special education services for those over three are key interventions. Early intensive communication and socialization-based interventions such as ABA (Advanced Behavioral Analyisi) and TEACHH Programs have been shown to be among the most effective interventions in improving the child's ability to develop language and communication skills and in helping with social integration.[19,20] Additionally, families should be linked up with support services such as the Autism Society.

The "M" stands for Monitor. Beyond monitoring, though, the primary care provider must also mentor the family through the subsequent learning and adjustment. After a definitive diagnosis of one of the Autistic Spectrum Disorders has been made, the primary care office must expand its role as a Medical Home not only to provide care but to insure access to primary and specialty care and care coordination. This should include monitoring of the child's overall health, immunizations and care of the typical illnesses and injuries of childhood. Because children with autism have similar health care needs as other children, the primary care provider must remain actively involved in the general pediatric care and not abrogate those responsibilities because of the diagnosis of autism. There should be continued surveillance for behaviors that might be related to or be outcomes of a child's autistic condition, such as altered eating, sleep patterns and toileting. Behavior problems may arise at times of physical stress, such as illness or the onset of puberty. Special consideration should be given to monitoring destructive, self-injurious or aggressive behavior. Progress in language development and behavior should be reassessed regularly..

After the diagnosis has been made, families may well return to their child's primary care provider for guidance about interventions, educational programs and treatments. Using a case-based learning approach, the pediatrician can become informed about Autistic Spectrum Disorders and be a valuable resource for the family. Consultation with a pediatric developmental and behavioral specialist is essential in the overall management of the care of the child with autism. Nevertheless, the primary care provider should be sufficiently versed in the care of children with autism to be able to answer basic questions. This may be particularly important in the areas of causation, intervention and those unproven treatments that promise improvement or even a cure. Recently the media has publicized a possible causative relationship between **measles, mumps and rubella (MMR)** immunization and autism. Despite several large studies failing to demonstrate any causative relationship, many in the public suspect a link.[21,22] The primary care provider can offer information, clarification and reassurance for families. Parents can feel confused when presented with unproven treatments that promise improvement or even cure; e.g., dietary

manipulation, therapeutic intervention or medications, such as intravenous administration of secretin or chelation therapy. Several studies have shown those treatments to be of no value in altering behavior or function of autistic children.[23,24] Yet some health care professionals support the use of secretin in the treatment of children with autism. The primary care pediatrician or family physician may be called upon to assist families in selecting interventions for their children.

The primary care provider may also need to advocate on behalf of the child and family with schools and health care plans. At other times, questions will arise about the transitions that occur in the lives of families with autistic children as indeed in all families of children with special needs. The first and most critical transition is at the time of diagnosis when parents must come to terms with their child's severe and potentially life-long disability. At the same time parents must face enrolling their child in an early intervention program. Though this is accompanied by the expectation of improvement, it is an additional confirmation of the child's disability. Later there will be the transition from early intervention at age three to a special education school program. At any time during childhood there may be crises over the child's behavior or developmental lags. In early adolescence the transition of educational, social and health care will begin, ending in the transfer of the young adult to adult care and service systems.[10] During each transition, the primary care provider may be asked to provide guidance.

The primary care pediatric provider, whether a pediatrician, family physician or nurse practitioner, plays a crucial, central and important role in the assessment of the child at risk of autism and in providing ongoing care after the diagnosis is made. The Academy of Pediatrics has recommended roles for the primary care provider in the diagnosis and management of children with Autistic Spectrum Disorders.[25] Important points for pediatric care providers are listed in Table 2.

Table 2. Fourteen Points for Providing a Medical Home for the Child with Autistic Spectrum Disorder and the Family

1. Be aware of the "Red Flags" for Austistic Spectrum Disorder.
2. Incorporate behavioral and developmental surveillance into health maintenance visits.
3. Use formal autism screening tools such as the **Checklist for Autism in Toddlers (CHAT)** or the **Pervasive Developmental Disorders Screening Test-II (PDDST-II)** when the possibility of Autistic Spectrum Disorders is suspected.
4. Refer to Early Intervention when any developmental risk is suspected.
5. Make an early referral to a pediatric behavior and developmental specialty team for a thorough diagnostic assessment when ASD is suspected.
6. Refer to a pediatric neurologist, geneticist and other specialists whose insights might be important in establishing causation.
7. Use case-based learning to improve knowledge and ability to provide care and support to the child and family.
8. After the diagnosis of Autistic Spectrum Disorders, put the family in contact with local and national autism support groups.
9. Assist the family of the autistic child to obtain emotional support, and refer to supportive and mental health services.
10. Partner with parents in a discussion of the diagnosis, treatment and intervention for the child, the parents and siblings.
11. After diagnosis, be vigilant for the developments of co-morbidities and specific sleep, eating and behavioral disorders, such as aggression or regression.
12. Advocate for the child and family with schools, service providers, state agencies and health insurers.
13. Be proactive at times of transition. Begin the planning process of transition to adult health care and service as early as 12 years of age with the transfer of care anticipated to take place as a young adult.
14. Provide a Medical Home with access to routine and coordinated care that is family-centered and culturally sensitive.

Though usually diagnosed in childhood and often considered a childhood condition, autism is a lifelong disorder with lifelong disabilities. The care of patients with autism needs to extend beyond childhood. Transition and transfer to adult care is an essential element for the autistic young adult. This will require improvements in training not only for providers of pediatric care but also for adult health care providers.[26]

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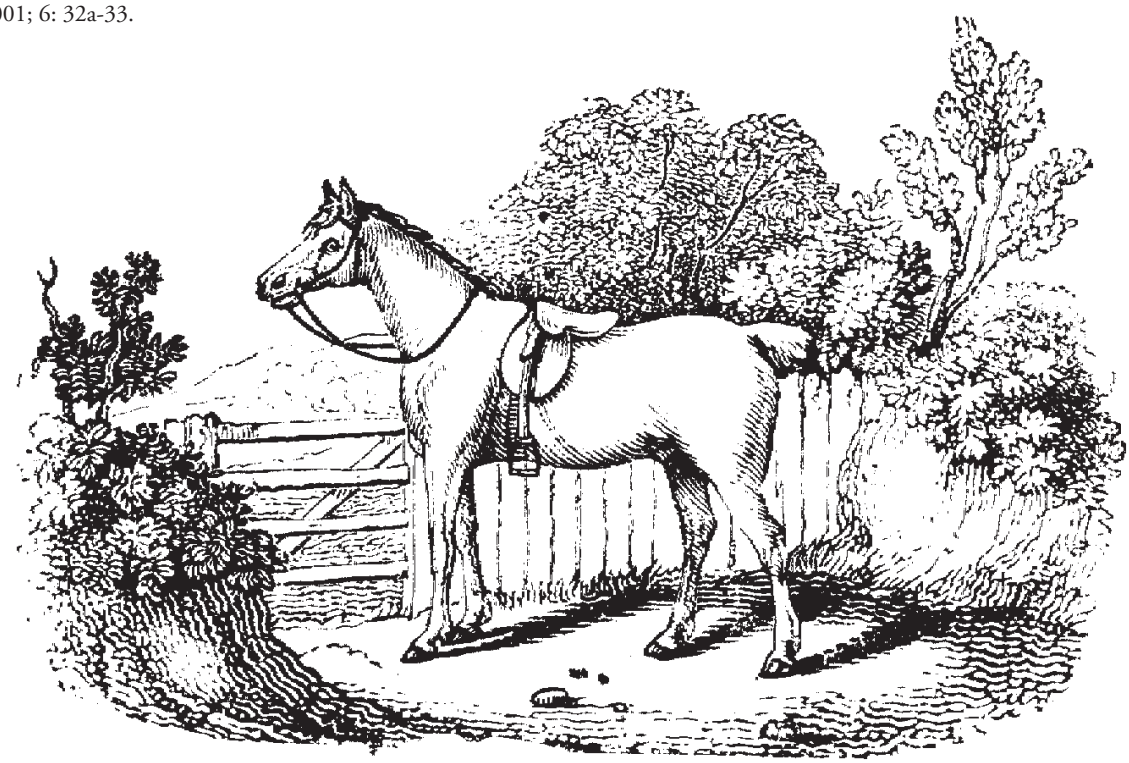
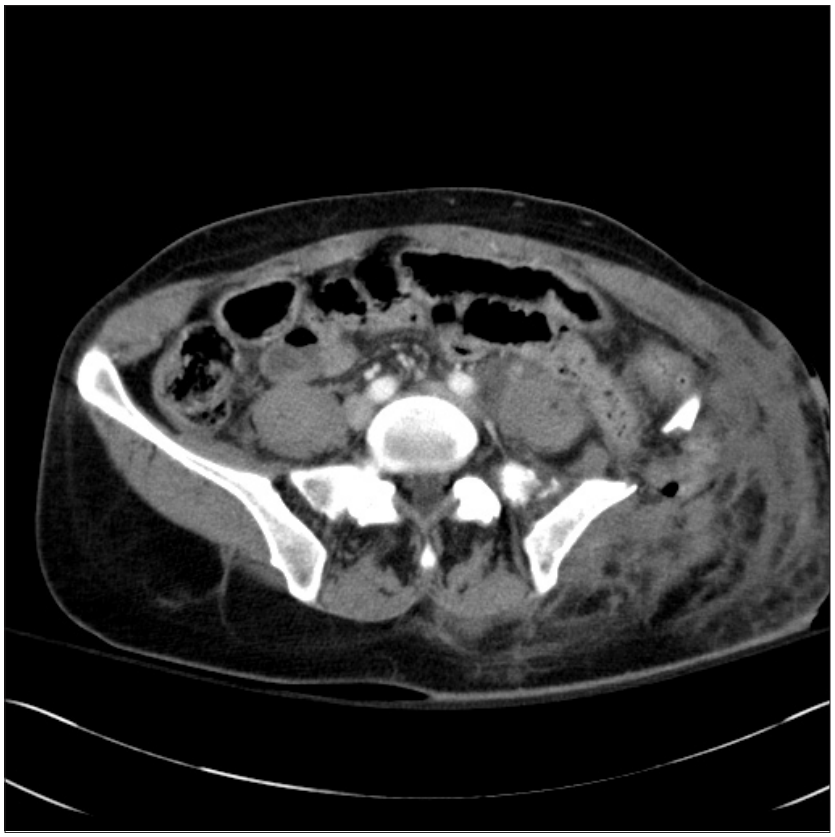


Images in Medicine

Bowel Entrapment in Comminuted Iliac Wing Fracture



A 56-year old female involved in a motor vehicle accident complained of left hip and back pain on arrival to the emergency department. Review of computed tomography (CT) prior to pelvic embolization by Interventional Radiology revealed an extruded loop of the bowel within a comminuted left iliac wing fractre (Figure 1). At emergent laparotomy a 4 cm longitudinal rent in the mid transverse colon and a 3.5 cm serosal tear in the sigmoid colon were repaired. The patient later underwent open reduction and internal fixation of the pelvic fractures.

Bowel entrapment is a rare complication of pelvic fracture with fewer than 5 cases reported in the literature. The diagnosis is often delayed and should be suspected in cases of prolonged ileus with pelvic fractures. Imaging findings of traumatically herniated or entrapped bowel is crucial because it maybe fatal if not promptly treated.[1]

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Performance of Rhode Island's Commercial Health Plans, 2003

Bruce Cryan, MBA, MS

Health plans differ in how they keep members well and how they care for them when they are ill. They also differ in how they provide access to care and deliver services. To consumers, the cost, quality, and access to care provided by a plan may affect their health. To employers, these same issues may influence worker absenteeism, productivity, and the company's personnel costs.

Of Rhode Island's commercially insured population, 88% receive their health coverage through four plans - Blue Cross and Blue Shield of RI, its wholly owned subsidiary BlueCHiP, United Healthcare of New England, and Blue Cross of Massachusetts. Information about how these plans perform is essential to determining if value is received from the premium dollars. In response to this need for information, the Rhode Island General Assembly passed the Health Care Accessibility and Quality Assurance Act in 1996. The Act instituted a program of health plan performance reporting in Rhode Island. Since that time, the state has become a national leader in this field.[1] The information presented here is derived from the program's most recent annual report on the performance of commercial health plans in the state.[2]

Methods

The Rhode Island Department of Health uses an annual survey to collect health plan data from three primary audited sources: Statutory financial filings, **Health Plan Employer Data and Information Set (HEDIS)** reports[3] and **Consumer Assessment of Health Plans (CAHPS)**[4] reports. This survey is supplemented by utilization review information also reported by the plans.

Thirty-six measures are collected, which fall into nine separate dimensions of performance (enrollment, finances, utilization, prevention, screening, treatment, access, satisfaction, and utilization review). To gauge performance, the measures are analyzed over time (i.e., trended) and compared to national and **New England (NE)** benchmarks.[5]

Results

Rhode Island's commercial health insurance market is concentrated in two carriers. Blue Cross and Blue Shield of RI, with its subsidiary BlueCHiP, has a market share of 64%, and United Healthcare of NE controls 18%. Blue Cross of Massachusetts has made some inroads, but its share remains in the single digits (7%). The remainder of the market (12%) consists of a number of smaller plans, none of which has more than 10,000 fully-insured RI members.

Average monthly health plan premiums in 2003 were 25% higher in RI than in the US ($248 versus $198), but 5% less than in NE ($248 versus $261). (Figure 1) RI plans spent 26% more on healthcare services than did plans nationally ($209 per member per month versus $166), and slightly less than regional plans ($209 versus $219). The higher expenditures for health care services may be partly due to Rhode Islanders' greater use of hospital services. The inpatient day utilization rate was significantly above both US and NE rates (11% and 21% higher, respectively), and utilization of hospital **emergency departments (EDs)** was 9% greater than the US rate (but comparable to the NE rate).

In addition, local plans incurred 22% more administrative expenses than US plans ($31.02 versus $25.51), but about the same as their NE counterparts ($31.02 versus $31.52). Statewide, health plan profitability peaked in 2003, with a $8.60 underwriting profit per member per month compared to a $6.89 profit nationally, and a $10.72 profit in NE.

Rhode Island health plans generally performed comparatively well on 20 clinical and access quality measures in 2003. (Table 1; see Reference 2 for full definitions of measures.) Overall, RI plans improved on eight

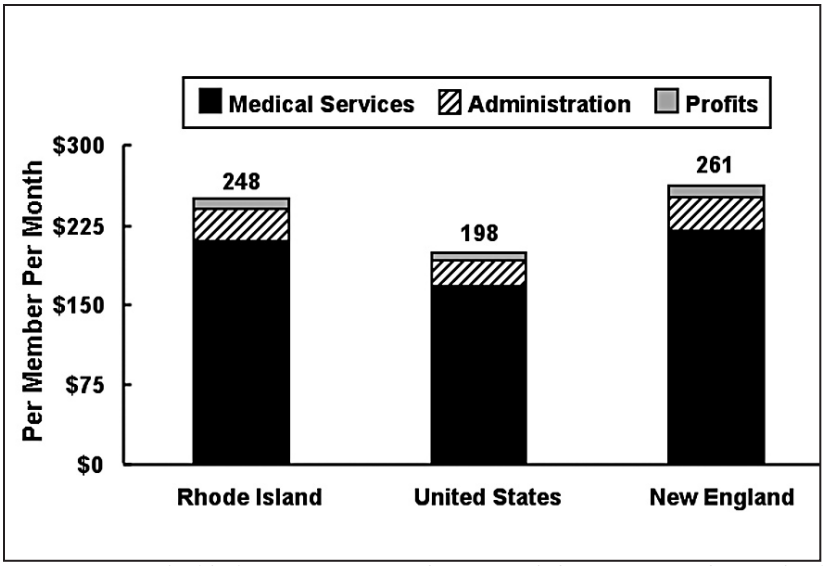

Figure 1. Average health plan premium per member per month, by component and geographical area, Rhode Island, United States, and New England, 2003.

Table 1.
Health Plan Performance Based on Clinical Measures,
Rhode Island (2002, 2003), New England (2003), and United States (2003).

Dimension/Measure	Rhode Island, 2003 Compared to -			
	Rhode Island, 2002		New England, 2003	United States, 2003
Prevention				
Childhood Immunization	=		=	+6%
Adolescent Immunization	+9%		=	+32%
Advising Smokers to Quit	+12%		=	+8%
Screening				
Breast Cancer Screening	=		=	=
Cervical Cancer Screening	=		=	=
Chlamydia Screening	+19%		-6%	+10%
Diabetes Care: Eye Exam Screening	=		-6%	+15%
Diabetes Care: HbA1c Tested	=		=	=
Treatment				
Controlling High Blood Pressure	=		+7%	+14%
Beta Blocker Treatment	=		=	=
Cholesterol Management	+5%		=	+5%
Diabetes Care: HbA1c Controlled	+10%		-11%	=
Antidepressant Medication Management	+19%		=	+43%
Access				
Follow-up for Mental Illness	+9%	-6%		=
Prenatal Care Access	=	-10%		=
Postpartum Care Access	=	-5%		=
Well Child Visits	=	=		+28%
Adolescent Well-Care Visits	=	+8%		+60%
Mental Health Access	=	+25%		+79%
Substance Abuse Access	+7%	+65%		+114%

Note: "=" indicates differences of no more than 5%.

measures (40%) and held steady on the remaining twelve measures (60%) when compared to 2002. In addition, on these 20 measures, RI surpassed the national benchmarks on twelve measures (60%), and was comparable to those benchmarks on the other eight measures (40%). [Note: Differences of less than 5% are not considered significant.]

Compared to the regional experience, RI plans did less well. On the 20 quality metrics, RI surpassed the New England benchmarks on four measures (20%), were comparable to those benchmarks on ten measures (50%), and fell below the benchmarks on 6 measures (30%).

Even though RI health plans' comparative performance was quite favorable, the absolute values on certain measures are a concern. An example is *Antidepressant Medical Management*, an 'effectiveness of care' treatment measure. "Effective" in this case means not that the underlying disease was cured, but that the treatment was "optimally" managed. RI's 2003 value improved by 19% from 2002, and was a full 43% higher than the US benchmark. Nevertheless, the absolute value for Rhode Island plans was only 29%, clearly leaving room for improvement.

Two-thirds of Rhode Islanders were satisfied with their health plans and four-fifths were satisfied with their healthcare. (Figure 2) RI's healthcare satisfaction rate was 6 percentage points higher than the national rate and similar to the regional rate. Rhode Islanders' satisfaction with their health plans was 4 percentage points higher than the national rate and also similar to the regional rate. Interestingly, regardless of geographic area, more members were satisfied with their healthcare services than with their health plans.

Discussion

Increasingly, the public, purchasers, providers, and policy makers are seeking meaningful information about health plans. Since 1998, the Department of Health has had formal data collection efforts to track and quantify the performance of this industry and has produced annual reports on the subject.[6]

With the small number of health plans in the state and the market dominance of Blue Cross and Blue Shield of RI, most Rhode Islanders have limited choice of carrier. The lack of selective contracting also means that most plans provide services through the same network of physicians, hospitals, and other providers.

Therefore, the real value in publishing performance information is less in aiding consumer choice of insurer and more in fostering accountability of the industry. Purchasers deserve to know how well the plans are performing and policy makers need empirical evidence to set effective policy. An added benefit of this effort is that the performance of health plans will improve if for no other reason than the results are publicly reported.

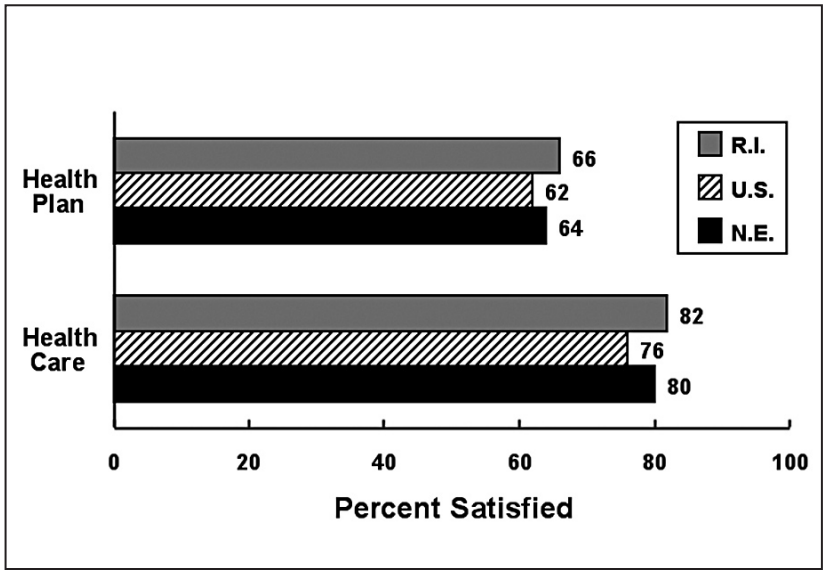

Figure 2. Health plan member satisfaction with health plan and health care, by geographic area, Rhode Island, United States, and New England, 2003.

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5. Financial benchmarks: National Association of Insurance Commissioners' Health database. All other benchmarks: Quality Compass of the National Committee for Quality Assurance.
6. Annual reports are available on the Performance Measurement and Reporting Program website: http://www.health.ri.gov/chic/performance/index.php.

QUALITY PARTNERS™ OF RHODE ISLAND

Doctor's Office Quality Focus on Information Technology

Physicians are challenged to provide better care for their patients while attempting to save time and money. Electronic health records (EHRs) can enhance access to patient information, decision support, and reference data, as well as decrease the likelihood of errors and improve patient-clinician communications. For most physicians, not currently using an EHR, the question is not *if,* but *when* and *how* to make the leap. Until now, a gap in educating physicians in this process has existed within the state.

Quality Partners of Rhode Island, in collaboration with the Rhode Island Medical Society, began the process of bridging this gap on March 5, 2005 when they co-sponsored a Health Information Technology Fair in Warwick. The agenda included presentations by national and local experts in Health IT as well as live demonstrations of EHR software. Dr. David Kibbe, Director of the Center for Health IT, American Academy of Family Physicians, gave the keynote speech, *"The Health IT Landscape: What Physician Leaders Need To Know"*, to an audience of over 150 attendees that included many local physicians.

The event was considered a kick-off for the Doctor's Office Quality – Information Technology (DOQ-IT) Project. This is a national program funded by the Centers for Medicare and Medicaid Services (CMS), designed to improve outcomes for patients with chronic illnesses by promoting the adoption of EHR systems and health information technology in physician practices. Quality Partners is the local quality improvement organization that will implement the program.

Although it has been demonstrated that EHRs can be major tools in addressing quality, cost, and time issues in clinical practice, effective implementation takes time, money and careful planning that many practices cannot afford. That is where Quality Partners and the DOQ-IT program can help.

Quality Partners will work free of charge to provide practices with assistance in workflow analysis and practice redesign, EHRs, selection and implementation and improved patient care management through EHR and quality improvement efforts. A roadmap for EHR adoption is outlined in the table below. For more information on DOQ-IT, contact Maureen Claflin, MSN, RN, Outpatient Project Manager at 528-3203. Recruitment for the project is currently underway.

	DOQ-IT EHR Road Map	
ADOPTION	1. Assess	2 Months
	2. Plan	2 Months
IMPLEMENTATION	3. Select	2 – 8 Months*
	4. Implement	4+ Months **
CARE MANAGEMENT	5. Evaluate	Ongoing
	6. Improvement	Ongoing

*Dependant upon duration for EHR selection by Physician Office
**EHR Implementation dependant upon installation complexity

This article was developed by Quality Partners of Rhode Island under contract with the Centers for Medicare and Medicaid Services, an agency of the U.S. Department of Health & Human Services. Contents do not necessarily reflect CMS policy.

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Letters To The Editor

To the Editor:

We were surprised to see the report published by Drs. Johnsingh and Snyder describing a case of **hemophagocytic lymphohistiocytosis (HLH)** in *Medicine & Health/Rhode Island*, August 2004 (volume 87, #8). This case is easily recognizable as a patient cared for by us and we certainly agree that it was a complicated case with many unusual features that warranted publication. Evidently, it is part of a series of cases seen by Brown residents that seems to straddle the formats of case discussion and case report. It is very distressing, however, that neither of the physicians claiming joint authorship of this article had any role in the actual care of the patient, nor did they discuss their intention of publishing it with us.

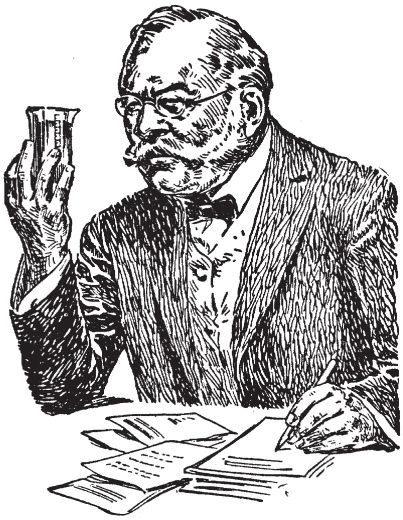
An enormous amount of effort went into establishing both the diagnosis and the treatment that is described in this report. This included an extensive literature review by us, discusions with national experts (Dr. Robert Arsceci and his colleagues at Johns Hopkins were extremely helpful) and extensive discussions with our pathologists, Dr. Lewis Glasser and Dr. Ronald Dellellis. In particular, the determined efforts of the pathologists to rapidly establish a diagnosis here were almost certainly life saving.

The authors of this case report acknowledge none of this. Some of the information is in fact not correct. We did not follow the 2004 HLH protocol as reported. HLH 2004 called for the initiation of cyclosporin, but because the patient was neutropenic, on steroids and had an established diagnosis of lymphoma, and we felt that cyclosporin was likely dangerous and unnecessary. The HLH 2004 protocol was drafted to address the familial form of disease and we felt that this was a different clinical situation. The favorable outcome here suggests that this is correct.

Whether the true issue here is authorship or simple basic courtesy to professional colleagues, this report does a disservice to the physicians in the community who actually made the diagnoses and established the treatment plan that is described here. We are very pleased to see physicians in training preparing difficult cases for publication so the lessons learned can be shared with the wider community in your journal. We hope that in the future a more thoughtful editorial policy will not trivialize the efforts of those of us for whom this is more than mere intellectual exercise.

On a happier note, we are pleased to report that this patient is entirely well after completing therapy for his lymphoma (eight cycles of CHOP) and is disease-free at nearly one year at last follow up (February, 2005). The underlying disorder, anaplastic large cell lymphoma, expressing ALK (anaplastic lymphoma kinase) has been found to have a five year disease-free survival of approximately 75-80% since it was identified as a distinct pathologic entity in the 1990s. Contrary to the suggestion in the article, lymphoma-associated HLH most likely has a prognosis in line with the underlying disease once the HLH itself is brought under control. Currently, it is probably much better than the median survival of 83 days that Johnsingh and Snyder cited from a previous treatment era. We suspect the prognosis here was also improved by the internet-facilitated access to critical information on the disease and treatment as well as the rapid and the outstanding laboratory support that is locally available.

Peter Rintels, MD
Sundarsesan Sambandam, MD



A Physician's Lexicon

The Materials of Medicine

A major component in the formal curriculum of Western medical schools prior to the 20th Century was an intensely pragmatic course called *Materia Medica* [medical materials]. The studied materials were those agents, sometimes called drugs, which were employed in the treatment of human disease. They might be simple or compounded; botanical [often called galenical], of animal origin or chemical [natural or synthetic].

The phrase, *Materia Medica*, was gradually supplanted by the word, *Therapeutics*, derived from a Greek word meaning "inclined to serve" or "be attentive to." Therapy once had a broader agenda but its meaning has now been narrowed to signify only that branch of medicine concerned with treatment. A remotely related Greek word, *theriac*, [literally, a wild beast] describes antidotes to poison. The word had been coined by Andromachus, a Crete physician, who had concocted an antidote for snake bites as well as for the bites of other feral creatures. A *mithridate* was a medication also designed to neutralize a poison. It was named for the somewhat paranoid king of Pontus, Mithradates [132 - 63 BC], who had undertaken extraordinary measures to protect himself against poisoning. His name was derived from Mithra, the ancient Persian god of light. *Alexipharmic* [Greek, meaning to ward off poison] is yet another term for an antidote. And the word, antidote, is similarly of Greek origin, the word meaning "given against."

By the middle decades of the 20th Century, medical schools designated this branch of medicine as Pharmacology, from the Greek word, *pharmicon*, meaning drugs. Much of the earlier classroom activity of pharmacology concerned itself with the technical preparation of various categories of medications.

A lotion, for example, was an aqueous solution used primarily for washing [from the Latin, *lotus*, to wash]. A potion, on the other hand, was any liquid medication to be taken orally [from the Latin, *potio,* to drink]. Cognate words include potable [suitable for drinking], symposium [literally a drinking together] and potomania [an older term for alcoholic frenzy].

An elixir, originally defined as a substance to prolong life, is from the Arabic, *al-iksir*, meaning the dry substance. A syrup, a thick and often sweet fluid, is also from an Arabic word, *sharab*, meaning a beverage. Cognate words include sherbet and sorbet.

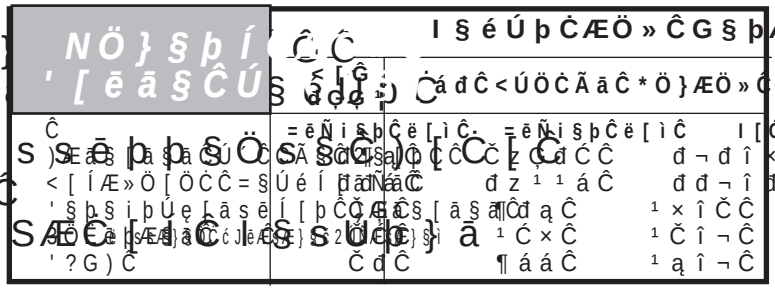
A tincture, an alcoholic solution of a medication, is derived from the Latin, *tingere*, meaning to dye or color. A liniment, defined as a liquid medication applied to the skin, is taken from the Latin, *linere*, meaning to anoint or smear. A balm, a fragrant and often resinous medication comes from the Latin, *balsamum*, meaning the gum of the balsam tree. The word, embalm, defines the process of preserving the dead body with resinous substances. And a decoction is a medication prepared by boiling [from the Latin, *decoctus*, meaning to boil down].

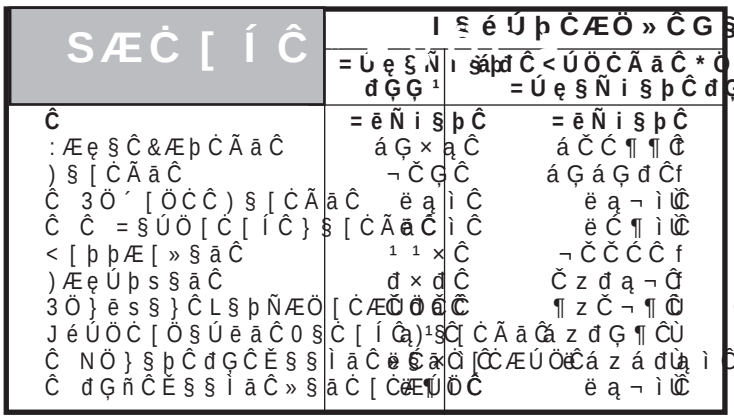
Stanley M. Aronson, MD

Rhode Island Department of Health
David Gifford, MD, MPH,
Acting Director of Health

Vital Statistics

Edited by Roberta A. Chevoya, State Registrar





(a) Causes of death statistics were derived from the underlying cause of death reported by physicians on death certificates.

(b) Rates per 100,000 estimated population of 1,069,725

(c) Years of Potential Life Lost (YPLL)

Note: Totals represent vital events which occurred in Rhode Island for the reporting periods listed above. Monthly provisional totals should be analyzed with caution because the numbers may be small and subject to seasonal variation.

* Rates per 1,000 estimated population # Rates per 1,000 live births

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Ninety Years Ago, May 1915

The Carlisle, PA, local medical association had published a list of names of patients "who were slow in making payments." One patient, who deemed the action libelous, brought suit against the reporting physician. A judge ruled: "There was no evidence of malice or any damage done." Since one or more Rhode Island District Societies promulgated such a list, the Editorial reported the decision.

A second Editorial reported that the Providence Medical Journal was on the abstract list of the Journal of the *American Medical Association*.

Ross McPherson, MD, in "Indications and Contraindications for Abdominal Caesarean [sic] Section," praised the procedure. "…one who sees this operation done in a skillful and successful manner is apt to query, Why is this not an ideal manner in which to have all babies enter the world?" But Dr. McPherson qualifies his enthusiasm: "There is a definite mortality rate to any laparotomy; small, it is true, in properly conducted cases, but still present in a much larger percent than in normal labor."

Frederic J. Farrell, MD, in "Observations on the Colloidal Gold Reaction," noted Zsigmondy's 1901 discovery "that proteins precipitated solutions of colloidal gold, and also that proteins in the presence of an electrolyte prevented this precipitation; whereas the electrolyte alone would precipitate the colloidal metal." In 1912 Lange noted the diagnostic impact of this discovery. Dr. Farrell examined the spinal fluid from 96 patients for dementia paralytica, cerebrospinal syphilis, and tabes dorsalis. He found the test "of value."

Fifty Years Ago, May 1955

William B. O'Brien, MD, Orlando Armada, MD, and Norman J. Wilson, MD, from the RI State Sanatorium, Wallum Lake, contributed "Modern Attack on Pulmonary Tuberculosis," focusing on thoracic surgery. They reviewed 411 patients with unilateral and 180 patients with bilateral surgeries, from 6 month to 7 and a half years-post surgery. "Our figures indicate that the prognosis for a given patient depends chiefly on the control of the open lesion rather than on the type of surgery." They concluded: "95% of all our living patients treated by various surgical methods are completely well."

Earle F. Kelly, MD, Hrad Zolmian, MD, and Banice Feinberg, MD, in "Treatment of Acute Infectious Meningitis in a General Hospital," recommended: "…any communicable disease should, and can, be handled effectively in a small general hospital like the Pawtucket Memorial."

One Editorial, "Looking Ahead in Mental Retardation," lauded RI Congressman Fogarty, chair (for the 5th time) of the House Committee responsible for the Health, Education and Welfare budget. The Congressman was instrumental in the inclusion of a new budget item - $750,000 "to initiate a program of research on mental retardation." The Editorial also noted that the RI General Assembly had passed a bill appropriating $400,000 to reimburse towns for half the cost of providing special facilities or transportation to existing facilities for retarded students in day school…" The Congressman also urged use of the term, "exceptional" children.

A second Editorial, on the results of vaccinating 1,830,000 schoolchildren, ages 6 to 8, in 44 states, for polio, reported the vaccine was "60-90% effective."

Philip Batchelder, MD, reviewed the book, *Of Smoking and Cancer: A Doctor's Report,* by Alton Ochsner, MD. The reviewer argued against the "crusader spirit," comparing the author to "zealous temperance workers of yesteryear." Although Dr. Batchelder accepted the facts and figures, he concluded, "I have not stopped smoking yet."

Twenty-Five Years Ago, May 1980

Arnold Porter, MD, in "Categorization of Hospitals in Rhode Island," advised: "Only Rhode Island Hospital can fulfill the requirements for a regional trauma center in Rhode Island."

Stanley M. Aronson, MD, in Dean's Message, commented on "Internships Obtained by the Brown Medical Students, class of 1980." Of 61 graduates, most intended to practice in the Mid-Atlantic states (23), with New England coming in second (13).

Robert E. Knisley, MD, Richard P. D'Amico, MD, and Joseph Di Benedetto, Jr., MD, in "Lennert's Lymphoma – Repot of a Case with Unusual Presentation and Therapeutic Implications," discussed the "unusual neoplasm characterized by high content of epithelioid histiocytes presenting as gastric lymphoma."

Constantine P. Pagonis, MD, discussed "Prednisone Therapy of Gold-Induced Thrombocytopenia in a Rheumatoid Arthritis Patient." The 67 year-old woman demonstrated the effectiveness of the regimen.

Reverend Joseph L. Lennon, OP, Vice-President for Community Affairs, Providence College, contributed "Authority, Freedom and the Teenager." He discussed the "cult of rebellion," urging teachers to act in *loco parentis*.

Forthcoming

Medicine & Health/Rhode Island

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Community Health

Guest Editor:

Vincent Mor, PhD

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