Minding Your P's and Q's

MENTAL RETARDATION: SEVERITY AND SURVIVAL

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Mental retardation is considered an *Axis II* diagnosis under the *DSM*, where "specific developmental disorders" are grouped along with disorders of personality. A diagnosis of mental retardation under the *DSM* (*Diagnostic and Statistical Manual of Mental Disorders* of the American Psychiatric Association) generally requires significantly subaverage intellectual function with concurrent deficits or impairments in adaptive functioning and onset before age 18. The *ICD-9* (*International Classification of Diseases*, 9th edition) uses the same numerical codes for mental retardation (317-319) that the *DSM* does, which basically classifies by **severity**.

Mental retardation is a phenotype of diverse etiology with no single natural history, and published mortality abstracts are few. Mental retardation was the subject of Abstract #215 in the 1976 *Medical Risks* book (Pacific State Hospital — U.S.), and was included as a category of mental disorders whose mortality was reported on in Abstract #1228 of the 1990 *Medical Risks* monograph (in-patient mental health data, Dutch experience). General information regarding long-term prognosis of mental retardation, by etiology, exists in various pediatric medical, genetics, and psychiatric texts, but such data is somewhat eclectic. Moreover, the etiologic approaches to prognosis are limited by the fact that the cause of mental retardation is idiopathic or indeterminate in as many as 40% of patients (see *Table 1*). Stratification by **severity** grouping instead of etiology offers an alternative way of assessing prognosis, but longitudinal studies of this type (at least ones of any significant size or duration) are rare. Eyman's data, therefore, are somewhat unique, and Singer's abstract in this issue of the *Journal* summarizes the mortality experience after 11 years of follow-up in Eyman's large subgroup of severely disabled persons (over 7,000 subjects).

The severity of mental retardation in Eyman's study was characterized by *severe or profound mental retardation*, tested or presumed, and by *immobility* and *incontinence*. Eyman used mental retardation criteria largely consistent with *DSM-III*, the version of the *DSM* newly in use at the time the study commenced (1980). (These criteria have remained essentially unchanged through *DSM-III-R* and the draft version of *DSM-IV*, and are themselves based on the classification scheme endorsed by the American Association on Mental Deficiency.) These criteria typically require: 1) significantly subaverage general intellectual functioning, 2) resulting in or associated with deficits or impairments in adaptive behavior, 3) with onset before age 18.

General intellectual functioning is defined as an IQ obtained by assessment with one or more individually administered intelligence tests. *Significantly subaverage* is defined as an IQ <70 (below two standard deviations from the average) on such tests. (IQ's below 70 have been further subclassified in ways such as those illustrated in *Table* 2.) *Adaptive behavior* is defined with respect to standards of personal independence and social responsibility expected of persons of comparable age and cultural group. While subject to some cultural variability — especially with regard to higher cognitive skills — these adaptive behaviors typically include age-appropriate locomotion, self-feeding, basic communication, and survival or self-protection skills.

Because of the third provision (onset before age 18), mental retardation is grouped in the DSM with "disorders usually first evident in infancy, childhood, or adolescence." But the diagnosis of mental retardation is age-dependent in more ways than the obvious one of requiring diagnosis before age 18. Except for the most severe forms of retardation that are evident from birth (often because of their association with other severe medical problems) or the ones that are acquired through a sentinel event (like meningitis or a head trauma), mental retardation has a latency to it — a period of time during which the impairments have not yet emerged as manifestly different from the rate or degree of development of other individuals of comparable age and circumstance. (*Retardation* itself, as the term suggests, involves deficits that are first or commonly perceived as a lag or delay in expected acquisition of skills or passage of milestones, as opposed to deficits which are frank and permanent.) The more severely retarded the individual, the earlier in life (closer to actual time of "onset") the diagnosis would be made. The milder the retardation, the more likely it will take formal testing or encounter with the progressive and cumulative demands of society and schooling to become appreciated.

True incidence rates for mental retardation (new cases/year) are difficult to measure, partly because time

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of onset and time of diagnosis may be years apart. Putting aside this counting issue, the estimated annual incidence rate for the U.S. is relatively low, at 54/100,000/year. Prevalence rates on the other hand are relatively high (one to three percent for all mental retardation, 0.5% for severe mental retardation). This is due to the long (on average) duration of disease that is characteristic of mental retardation. In a stable population, incidence and prevalence are related by duration of disease (P = I x d), with disease-duration being either time from onset until death-*from*-disease, or time from onset until death-*with*-disease. Overall, if mental retardation has an incidence of 54/100,000/year, and a prevalence of 25/1,000 (2.5%) then the average duration-of-disease for mental retardation would be about 46 years. This long average duration is one of the reasons that mental retardation (all severities combined) is such a significant contributor to disability in the U.S., being the leading cause of activity limitation, and the ninth leading cause of needing help in basic life activities (all ages combined).

But the differential effects of severity on incidence, prevalence, and duration are important to appreciate. The incidence of mental retardation is highest at school ages, but this is largely a result of individuals with mild mental retardation, discovered through testing or adaptation difficulties during school years. These individuals have similar mortality rates and patterns to the general population, and hence continue to contribute to prevalence figures unless they acquire a degree of adaptation by adult life that drops them from the rolls of the retarded, or otherwise become socially assimilated and lost to follow-up or tracking. Severe retardation has a higher incidence in early childhood, but also has a higher mortality rate due to the high frequency of associated physical disorders and complications. So, severe mental retardation has a skew in both incidence and prevalence, and a correspondingly shorter "duration" of disease.

The incidence rate for mental retardation for the U.S. (all severities combined) is 54/100,000 per year. The 0-5, 6-12, and 13-18 year-old age-groups have incidence rates of about 10,37,and 7 per 100,000/year respectively. Of these, severe retardation accounts for about 9, 5, and 2 per 100,000/year respectively.

In 1973, the IQ cut-off point for mental retardation in the U.S. was made its current two standard deviations below normal for the population. Prior to this, the cut-off was one standard deviation. The change instantly eliminated a category listed as "borderline" mental retardation, and created today's categories (mild, moderate, severe, and profound). It also reduced the prevalence of mental retardation by 80% in one fell swoop, establishing IQs <70 as the threshold for "retarded."

In Eyman's study, the study subgroups were all *severely* retarded. They differed from the entire California registry (mental retardation, all severities combined) in the following ways:

	Entire Registry	Severely Retarded Subgroups
Mean-Age	17 Years	7 Years
Immobility	<23%	100%
Not toilet trained	29%	100%
Tube-feeding required	2%	100%
Severe retardation	43%	100%
Cerebral palsy	44%	74%
Seizures	42%	67%
Other major medical condition	10%	60%

Singer's abstract gives the long-term survival results of Eyman's subgroups 1,5 and 6 (*Table 3* shows the eight original subgroups). Singer has calculated comparative mortality in terms of mortality ratios (M.R.) and excess death rates (E.D.R). Eyman also provided information in terms of median survival (see *Table 4* for summarization). It is worth keeping in mind that the mean-age of each of Eyman's severely-retarded subgroups is overall quite young — ranging from 6.5 (subgroup 1) to 8.2 (subgroup 3). When subgroups 1 through 6 are age-stratified, the less than one year-old age-group has markedly curtailed life expectancies in virtually all subgroups. Possession of a few basic skills, like ability to roll (subgroups 5 and 6), and ability to feed oneself (subgroups 3, 4, and 5) were associated with the best survivals among this study of immobile, incontinent, severely retarded individuals. And the higher the attained age, the better the survival prospects. But as Singer's *comparative* mortality shows, mortality ratios are still astronomical, across all ages and subgroups out to 10 years follow-up.

The primary assessment tools for mental retardation are IQ scores and ADL-like basic ("survival") skills. The history of mental retardation has reflected the evolution of both medical diagnostics (power to differentiate types and severity of clinical conditions) *and* social awareness and attitudes about individuals who are *different*. The distinction between mental retardation and mental illness, for instance, is a fairly recent historical development (18th century), prior to which, mental institutions would often hospitalize the two indiscriminately. Degrees of severity of mental retardation came

to be recognized at about the same time, and terms were developed to attempt to describe them. Such terms as *idiot* (unique or peculiar to oneself), *imbecile* (without support, feeble of mind), and *moron* (foolish, uncomprehending) once had technical scientific meanings. But societal misunderstanding and caricature has led to sadly prejudicial and pejorative modern connotations. While IQ scores have largely replaced age-comparison (e.g., "mentally equivalent to a four-year-old"), scholastic potential (e.g., "trainable") and general descriptors (e.g., "moronic"), such IQ scores can be greeted with similar societal non-acceptance in a highly-competitive, grade-oriented, performance-based culture. But, perfect or not, IQ testing has provided a form of standardization in measurement not available before this century, and classification of mental retardation by severity has proven prognostically useful.

Figure 1
Study Group Formation

155,851	persons who received services from California Department of Developmental Services, between 1/80 and 3/91
	and
128,248	were followed for at least two years
	and
54,761	were severely or profoundly retarded
	and
7,228	had no improvement in basic movement or feeding skills during study period, and were traceable for outcome purposes.

(These 7,228 subjects were stratified to eight subgroups, as depicted in *Table 3*.. Study was a unizonal, multiserial, unisecular one.)

Table 1
Etiology of Mental Retardation

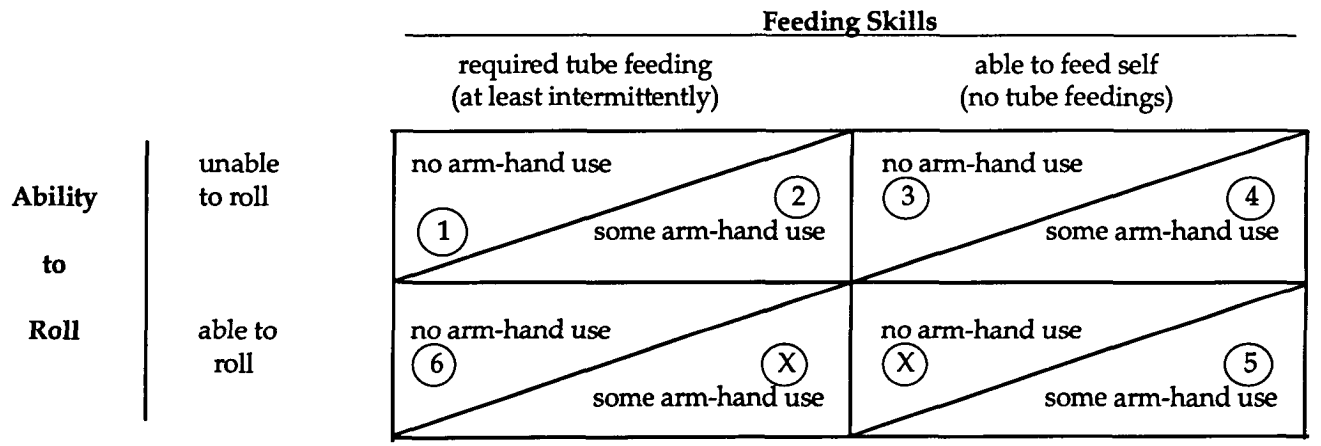
Cause	Estimated %
1) Hereditary (e.g. Mendelian or cytogenetic)	5%
2) Prenatal or embryonic damage	25-30%
3) Perinatal complications	10%
4) Acquired in infancy or childhood (e.g., infection, trauma, lead poisoning)	5%
5) Psychosocial factors and environmental deprivations	15-20%
6) Idiopathic or indeterminate	30-40%

Table 2
Ways in Which Severity of Mental Retardation Has been Categorized in the Past

		Historical Expressions			IQ-based
DSM3/ICD9	Descriptive Term	Esquirol/ Goddard Term	Schooling Implication	Abilities Comparable to Child of Age	Binet/Simons IQ Score
317	Mildly Retarded	moron	educable	12	50-70
318.0	Moderately Retarded	imbecile	trainable	4	35-49
318.1	Severely Retarded	idiot	non-trainable	2	20-34
318.2	Profoundly Retarded	idiot	non-trainable	<2	<20

Table 3
Mental Retardation Subgroups

(The study's subgroups are numbered 1-6; two of the 8 originally intended subgroups had insufficient numbers to include in the study, and are denoted by x.)



(All study subjects were also severely or profoundly retarded, immobile [unable to walk, crawl, creep, or scoot] & incontinent.)

Table 4
Median Survival by Subgroup

(All subgroups are comprised of severely or profoundly retarded persons with immobility and incontinence.)

Subgroup	≤1 Year	1-15 Years	16-49 Years
1	0.9	4.8	10.4
2	1.4	5.3	(n=26)
3	1.2	5.7	10.4
4	3.2	10.0	(60%)
5	(70%)	(70%)	(80%)
6	8.4	10.9	10.9

(where a % is given, median survival was not yet reached, and the% shown is the % still alive at end of FU. For subgroup 2, the 16-and-up age-group had such a small number of entrants that the authors did not consider median survival to be meaningful to report.)

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