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Intrauterine Rubella, Head Size, and Intellect

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ABSTRACT. The assumption that congenital rubella is commonly associated with microcephaly and mental retardation was examined. Among a rubella clinic population of 111 children, 92 children had vision sufficient to allow testing by the Leiter International Scale. The mean IQ for this group was 99.46 (SD, 19.5). Head circumference in this group correlated well with stature but poorly with intellect. The authors conclude that children with intrauterine rubella should be viewed as small children rather than children with small heads and that such children are poorly served if mental subnormality is assumed without careful study. *Pediatrics;* 55:797, 1975, RUBELLA, HEAD CIRCUMFERENCE, INTELLIGENCE.

Evidence abounds for the multiple handicaps of children afflicted with intrauterine rubella. Problems most commonly reported include auditory and visual impairment, congenital heart disease, small stature, and a variety of less common afflictions such as cerebral palsy, dermatoglyphic anomalies, genital defects, and diabetes mellitus. In addition, mental retardation and microcephaly are also believed to be common. In nearly every standard work on pediatrics or on mental retardation, one finds the implied or expressed statement that mental deficiency and microcephaly are common residua of prenatal rubella. In this paper we will examine this assumption.

While following a group of children with sensory handicaps in a multidiscipline clinic, it was our impression that although the children with intrauterine rubella might appear functionally delayed in motor, language, and adaptive skills, they seemed to differ from children with developmental delay due to other causes. First, many of the children with rubella appeared to "improve" with increasing age (causing frequent changes in classification). Secondly, there appeared to be a common tendency for the staff to "underestimate" their potential in contrast to the almost universal attitude shown toward other handicapped children in whom labeling is avoided and every benefit of doubt offered.

We were also impressed by the wide range of reported incidence of mental retardation among children affected by rubella[1-5] (Table I).

Chess and her co-workers[5] in their book on psychiatric disorders in children with rubella syndrome review the literature on the measurement of intellect in these children and conclude that mental deficiency exists in about one third of the children coming to the attention of centers. Similar incidence is reported by Cooper and Krugman[1] and by Gumpel *et al.*[3]

Sheridan, on the other hand, in the most systematic effort to date, was able to obtain intelligence test scores on 191 British children with evidence of prenatal rubella infection.[4] She reported that the mean IQ score was not only

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TABLE I

INCIDENCE OF "MENTAL RETARDATION" REPORTED IN RUBELLA SYNDROME

Author	*Year*	*Diagnosis**
Cooper & Krugman[1]	1967	35%
Menser *et al.*[2]	1967	10%
Gumpel *et al.*[3]	1971	37%
Sheridan[4]	1964	7%
Chess *et al.*[5]	1971	37%

*Developmental delay; retardation

TABLE II

DEFECTS OBSERVED IN RUBELLA SYNDROME (111 CHILDREN BORN BETWEEN 1964 AND 1966)

Defect	*No.*	*%*
Heart defect	50	46
Ocular defect		
Cataract/glaucoma	31	28
Retinopathy	92	83
Severe visual disability	14	13
Hearing disability	95	86
Extended syndrome/ encephalopathy	25	23
Low birthweight	60	54
Stature 2 SD or more below average	49	44

average but that the distribution approximated normality. Menser *et al.*,[2] in a 25-year follow-up on Gregg's original cases, reported a similarly low incidence of mental retardation in their group.

None of the reports made a distinction between developmental problems resulting from sensory deficit and innate intellectual deficiency. Few investigators have made a systematic effort to determine if these children are, in fact, intellectually deficient as clinical examination would suggest or to test in a situation in which the children were not penalized by their communication problems with vision and hearing.

Similarly, studies on groups of mentally retarded children[6,7] would tend to support the common teaching that associates "microcephaly" (head size more than 2 SD below the mean) with intellectual deficit. On the other hand, Martin[8] observed children with normal intellect and small heads and urged caution when using this measurement or drawing conclusions from it. No systematic data are available on a population of children with prenatal rubella to document the frequently reported description of microcephaly in this group or to correlate head size and intellect.

METHOD

Children studied were selected from a group with physical and sensory handicaps being followed in a multidiscipline clinic held at the Crippled Children's Division of the University of Oregon Medical School. A group of 111 children born between 1964 and 1966 had serologic or virologic evidence of rubella or had a prenatal history of rubella plus two or more manifestations of the postnatal syndrome (Table II). These children represent 2.0/1,000 live births during the period covered and it is believed that it includes most of the seriously affected children in the state who survived the epidemic. Of this group, 16 had visual and auditory handicaps sufficiently severe to prevent accurate psychometric measurement by standard testing instruments. Severe behavior problems existed in an additional three children who could not be properly examined. Data from the remaining 92 children were included in the calculations and constitute the subject of this report.

Each child underwent serial complete evaluations. Anthropomorphic measurements were obtained under controlled conditions.

To reduce bias introduced by language deficit in the deaf children, the Arthur modification of the Leiter International Scale[9] was offered at least once to each child. This test uses no spoken language and was developed originally to avoid language and cultural barriers. It has been found to be useful in the testing of the deaf and results have been found to correlate well with other testing instruments. Confirmatory measurements using the performance sections of the WISC[10] and Stanford-Binet[11] were obtained on a number of the children studied and found to be consistent. All children were tested between 4 and 6 years of age. The mean age at the time of testing was 57 months (SD, 9.2 months).

While standardized instruments for testing intelligence of blind children do exist, there are, unfortunately, no good instruments to use with children who are both blind and deaf. The elimination of this group, while small in number, admittedly introduces a bias into the present results. The spectrum of defects seen in these 16 children was similar to those in the tested group. Means for stature and head size did not differ significantly between the group eliminated and those evaluated by standardized tests for intelligence ($t = \pm 1.67$).

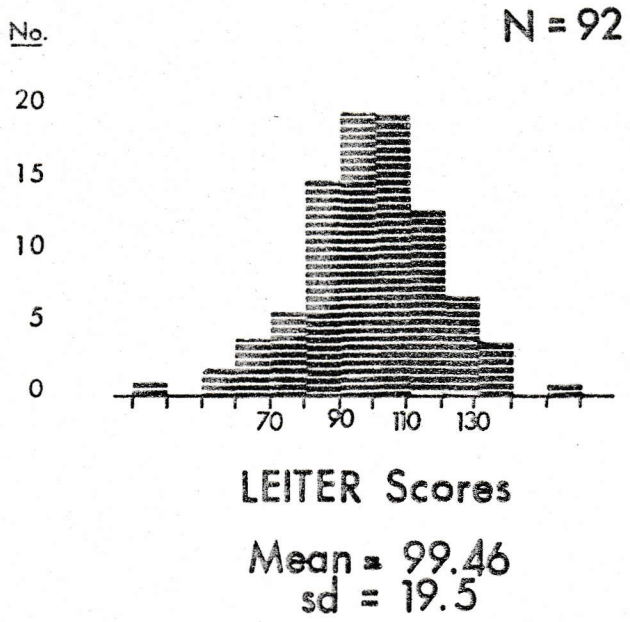

FIG. 1. Distribution of intelligence in rubella syndrome.

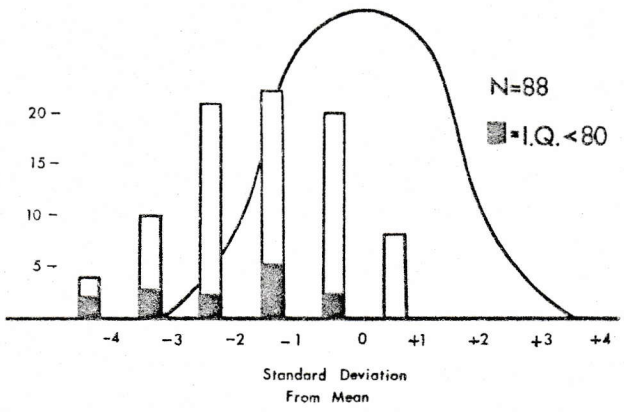

FIG. 2. Head circumference in rubella syndrome.

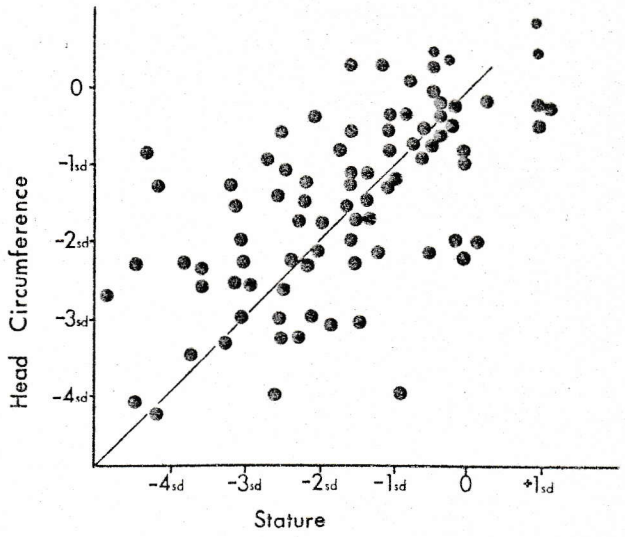

FIG. 3. Stature in rubella syndrome.

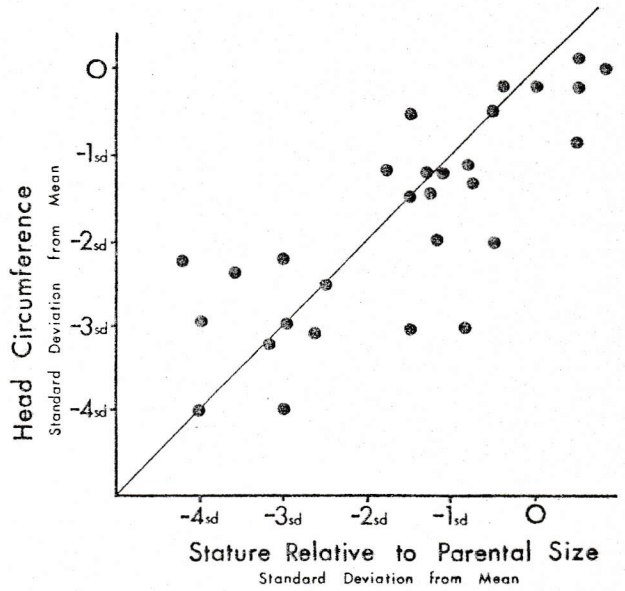

FIG. 4. Stature relative to parental size in rubella syndrome.

RESULTS AND COMMENTS

With the children with serious visual handicaps excluded, Figure 1 demonstrates the distribution of intellectual achievement as measured by the Leiter International Scale. As previously reported,[12] the mean IQ for this group was 99.46 (SD, 19.5; range, 63 to 153). The distribution curve is comparable to a sample of "normal" children.

Figure 2 demonstrates the distribution of head circumference of 88 of the 93 children on whom adequate data were available. It can be seen that, when plotted against published norms,[13] the occipitofrontal circumference for this group is smaller than for the general population. Though the numbers are small, it can also be seen, however, that children with measured IQ less than 80 are fairly evenly distributed through the population and there appears to be little correlation between intelligence and head size. In an attempt to determine whether small head size was an independent feature of the intrauterine rubella syndrome, a plot of head circumference

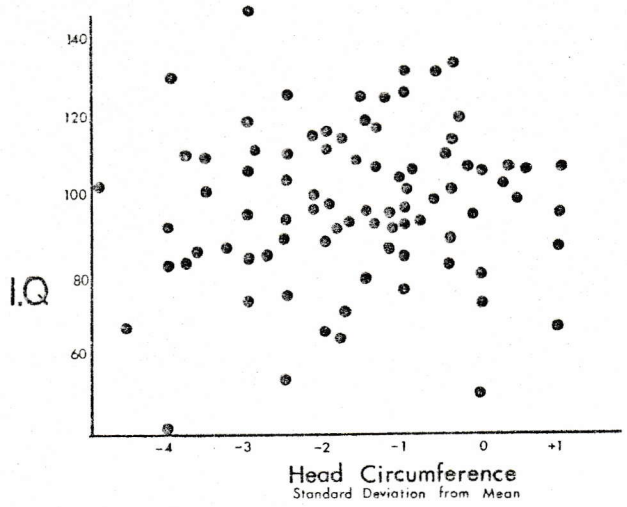

FIG. 5. Scattergram showing head circumference and IQ.

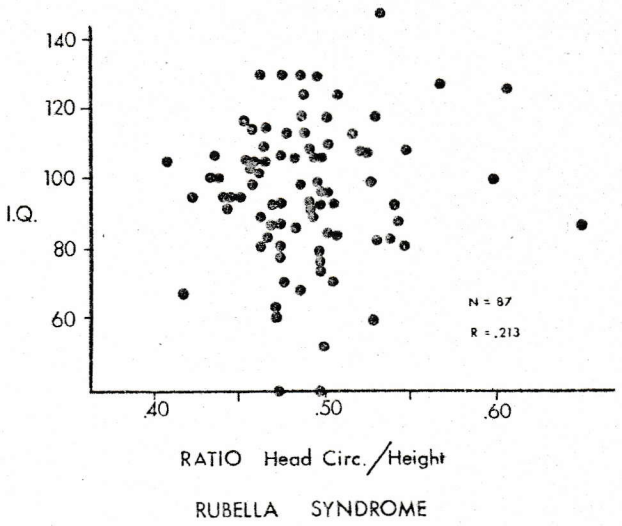

FIG. 6. Ratio of head circumference to height.

against stature (Figure 3) showed a remarkably good correlation ($r = .568$; $P < .001$).

In a smaller sample for whom parental data were available, head size was plotted against "expected" height relative to mean parental stature.[14] In this situation, the correlation was equally high ($r = 0.599$; $P < .01$) (Fig. 4).

Of greater significance, perhaps, is the distribution of head size relative to measured intellect. A plot of head size against IQ as measured by the Leiter scale (Fig. 5) shows very poor correlation ($r = 0.22$). If one postulates that those children with heads relatively small for body size will be more likely to demonstrate retardation, it is possible that such a correlation would not be shown in the previous calculation. We therefore compared intellect with a ratio of head size to total height. This ratio, though of little value when applied to all children, has a relatively narrow range (0.45 to 0.55) when calculated from standard data for children 3 to 6 years of age. Figure 6 shows such a plot. A statistically significant correlation between low intellect and small head size can be demonstrated in this way, but only barely at the 5% level of confidence ($r = 2.13$; $P = < .05 > .01$).

Despite the bias introduced by the exclusion of children with severe visual handicap, we believe these data tend to confirm the more optimistic prognostic views of Sheridan[4] and Menser *et al.*[2] regarding the intellectual potential of children with intrauterine rubella syndrome surviving the neonatal period. In addition, the results would suggest that the "microcephaly" which is commonly seen and described in these children appears to reflect small stature and that children afflicted with prenatal rubella should more appropriately be viewed as small children rather than children with small heads. They should, at least, not be indiscriminately grouped with mentally retarded children with microcephaly due to other etiologic factors.

The authors emphasize that they wish in no way to minimize the severity and lifelong tragedy of the handicaps associated with intrauterine rubella. Children with deafness, visual defect, or cerebral palsy *are* frequently delayed functionally and educationally handicapped. We believe, however, that unwarranted and unproven assumptions of intellectual subnormality are commonly being made in children with rubella syndrome who are already handicapped by sensory or motor defect. Such assumptions can in no way assist a child to achieve his or her greatest potential and, as pointed out by Rosenthal and Jacobson,[15] may lead to self-fulfilling prophecy.

REFERENCES

1. Cooper, L. Z., and Krugman, S.: Clinical manifestations of postnatal and congenital rubella. Arch. Ophthalmol., 77:434, 1967.
2. Menser, M. A., Dods, L., and Harley, J. D.: A twenty-five-year follow-up of congenital rubella. Lancet, 2:1347, 1967.
3. Gumpel, S. M., Hayes, K., and Dudgeon, J. A.: Congenital perceptive deafness: Role of intrauterine rubella. Br. Med. J., 2:300, 1971.

4. Sheridan, M. D.: Final report of a prospective study of children whose mothers had rubella in early pregnancy. Br. Med. J., 2:536, 1964.
5. Chess, S., Korn, S. J., and Fernandez, P. B.: Psychiatric Disorders of Children with Congenital Rubella. New York: Brunner/Hazel, 1971.
6. Payor, H. B., and Thelander, H.: Abnormally small head size and intellect in children. J. Pediatr., 73:593, 1968.
7. Nelson, K. B., and Deutschberger, J.: Head size at one year as a predictor of four-year I.Q. Dev. Med. Child Neurol., 12:487, 1970.
8. Martin, H. P.: Microcephaly and mental retardation. Am. J. Dis. Child., 119:128, 1970.
9. Arthur, G.: The Arthur adaptation of the Leiter International Performance Scale. J. Clin. Psychol., 5:345, 1949.
10. Wechsler, D.: The Wechsler Intelligence Scale for Children, manual. New York: The Psychological Corporation, 1949.
11. Terman, L. M., and Merrill, M.: Stanford-Binet Intelligence Scale, manual for the 3rd revision form L-M. Boston: Houghton Mifflin, 1960.
12. Dodrill, C. B., Macfarlane, D. W., and Boyd, R.: The effects of intrauterine rubella and its consequent physical symptoms upon intellectual abilities. J. Consult. Clin. Psychol., 42:251, 1974.
13. Nellhaus, G.: Composite international and interracial graphs. *Pediatrics*, 41:106, 1968.
14. Tanner, J. M., Goldstein, H., and Whitehouse, R. H.: Standards for children's height at ages 2-9 years allowing for height of parents. Arch. Dis. Child., 45:755, 1970.
15. Rosenthal, R., and Jacobson, L.: Pygmalion in the Classroom. New York: Holt, Rinehart & Winston, Inc., 1968.