

The next major agent in terms of environmental insult is infection. Here, too, there is a large bibliography in the literature but little precise information. There are only three diseases known to produce congenital defects. The first is rubella, about which we know more than other diseases. The second is toxoplasmosis, which is rare. The third, also rare, is salivary gland disease, sometimes called cytomegalic virus disease. The second and third of these diseases produce hydrocephalus, retinal calcification, and mental retardation. We do not understand the mechanisms involved, nor do we know why some women harboring these diseases at the right time produce normal infants and others produce abnormal infants.

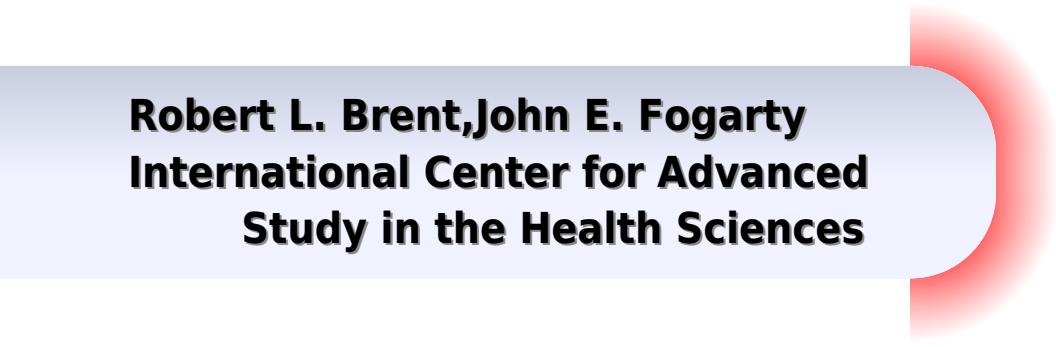
In the spring of 1964 a major German measles epidemic occurred in the New York area and other parts of this country. Whereas normally in New York about 2,000 to 3,000 cases of German measles are reported annually, more than 30,000 cases were reported in 1964 (and many others went unreported). In view of this situation it was anticipated that a large number of babies would be exposed to rubella in utero.

As expected, there was a high incidence of deafness and mental retardation in the newborn. The typical rubella triad of cardiac anomalies and cataracts was also encountered. What was not expected, however, was the marked increase of a serious congenital syndrome. Many babies were born underweight and rapidly developed thrombocytopenic purpura with hemorrhage. This syndrome has not been well documented previously. Many of these children died of hemorrhage, and there was a large variety of other manifestations. These data are reported by Krugman and Cooper at New York University, where, at the time of writing, the cases of almost 200 infants who manifested this syndrome were being followed. Of the 200 infants, the laboratory was able to isolate 92 cultures of rubella virus.

This syndrome was a relatively new finding. These babies had been infected by the rubella virus in the first trimester of pregnancy, six months before they were born. At birth they were still shedding the virus despite the fact that the mother and the infant did have hemagglutinating antibody in their circulation. There were 70 babies with the purpura syndrome, from which the virus was recovered in 58. Hepatomegaly, splenomegaly, and congenital heart disease were other major manifestations. Eye lesions (cataracts, cloudy cornea, or glaucoma), adenopathy, and bone lesions were also noted. The bone lesions came as a surprise, inasmuch as they had not been reported before. The lesions were almost exclusively in the distal portions of the femur and occasionally in the proximal portion of the tibia. Areas of radiolucency, an apparent

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**Robert L. Brent, John E. Fogarty
International Center for Advanced
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