

Harlequin Ichthyosis Among the Navajo: Counseling Issues

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Harlequin Ichthyosis is a rare, fatal congenital disorder of keratinization characterized by thickened, scale-like plaques of skin with a diamond configuration. Autosomal recessive inheritance has been established, and prenatal diagnosis for this disorder remains controversial. Five infants with this disorder were born among approximately 25,000 Navajo women who delivered in Gallup, New Mexico from 1970 to 1989. The incidence of 1 in 5000 among the Navajo in Gallup is high compared to previous reports in other populations. Two families with affected infants were seen in a genetics outreach clinic. Two important counseling issues were raised: (1) reluctance to discuss the possibility of recurrence for fear of affecting the outcome, and (2) feelings of guilt caused by cultural beliefs which attributed the birth defect to paternal behavior during pregnancy. Such issues are encountered in many traditional cultures, and they can be addressed by tailoring the timing and content of counseling.

KEY WORDS: Harlequin Ichthyosis; genetic counseling; Navajo culture; prenatal diagnosis; guilt.

INTRODUCTION

The Navajo people, or *Dineh*, as they call themselves, live in an area the size of West Virginia in the high desert of the Southwest. The reservation extends across northeastern Arizona to northwestern New Mexico,

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and into southern Utah. The Navajo now number approximately 200,000 (Indian Health Service, 1991). In a time of striving for economic self-sufficiency, the Navajo still strongly adhere to their cultural, social, and traditional values. Many live in traditional dwellings called *hogans*, eight-sided log homes with earthen floors and wood stoves at the center. Rug weaving, sheepherding, and silversmithing are some of their forms of livelihood.

Access to medical care is limited by their lifestyle. Many live miles from the nearest clinic or hospital, along dirt roads, and not all have vehicles for transportation. The Gallup Indian Medical Center (G.I.M.C.), the Public Health Service hospital on the eastern side of the reservation, serves a population of 38,000 Native Americans. Nearly all of the approximately 1200 annual births are to Native Americans. To help meet the needs of this large rural population, a genetic counseling outreach clinic was established in 1988 for the G.I.M.C. Department of Obstetrics, with support from the University of New Mexico Department of Obstetrics and Gynecology.

The first step in establishing this clinic was providing better access to genetic counseling services; the second step was understanding the traditional beliefs of a culturally distinct population. Navajo beliefs regarding pregnancy and childbirth include traditions integral to remaining in harmony with their natural, spiritual, and cultural world. A person may fall out of harmony with nature by his actions, omission or commission, resulting in sickness for himself or his family. Disharmony is a consequence of breaking taboos in everyday life. For example, tying knots while weaving rugs would result in a difficult childbirth. If a woman used a six-pronged weaving comb, the child would be born with six toes (Waxman, 1990).

Similar taboos have been described with much consistency from one end of the Navajo reservation to the other, including avoidance of dead animals or funerals during pregnancy. By appreciating cultural differences, some barriers to genetic counseling are more easily overcome.

HARLEQUIN ICHTHYOSIS: INCIDENCE DATA AND MEDICAL GENETIC ISSUES

The birth of two infants with Harlequin Ichthyosis within 4 months in unrelated families prompted a review of death records at G.I.M.C. from 1970 to 1989. In the review, a total of five documented affected live births were found. Each affected infant was from a different Navajo family. Thus, the incidence of Harlequin Ichthyosis was approximately 1 in 5000 at G.I.M.C., a much higher rate than expected for this usually rare genetic