OSTEOGENESIS IMPERFECTA: PREGNANCY CHARACTERISTICS, MODE OF DELIVERY, AND NEONATAL OUTCOME.

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From a database of 1016 individuals in whom OI was confirmed by biochemical studies of cultured fibroblasts or chorionic villus mesenchymal cells, collected between 1987 and 1994, we selected 547 that involved young children or pregnancies. Of the 547, obstetric information was available for 296. The studies were derived from electively terminated pregnancies or early miscarriages in 129 instances, leaving 167 affected individuals for whom delivery information was available. The obstetrical records were reviewed for prenatal detection, the method of delivery, the presence of prenatal and/or postnatal fractures, and, for the lethal forms, the length of survival.

In this cohort of pregnancies there were 29 with OI type I, 50 with OI type II, 5 with OI type II/III, 14 with OI type III, 37 with OI type III/IV, and 32 with OI type IV. Within this group the diagnosis of OI was suspected prenatally as a result of ultrasound studies in 39 (23%). Furthermore, the rate of breech presentation at term (44/120, 37% of infants delivered at term) was significantly higher than the expected 3-4% rate for the general population. The rate of cesarean delivery was 54% (90/167). Among those delivered by cesarean section the diagnosis of OI was known prior to delivery in 14 (16%). Although knowledge of a diagnosis of OI may have played some role in the choice of mode of delivery, it did not seem to be the major determinant of cesarean delivery. Instead, breech presentation (39/90 delivered by cesarean section) appeared to a better predictor for the choice of non-vaginal delivery. Cesarean delivery affected neither the fracture rate per infant nor the number of infants with fractures among those with the non-lethal OI phenotypes. Furthermore, cesarean delivery did not prolong the survival of infants with the lethal form of OI.

Thus although cesarean delivery is often recommended when faced with a pregnancy in which OI is known or suspected it does not appear to reduce fracture rate in all infants or to increase survival among infants with OI type II. The usual reasons to choose this mode of delivery are similar to those for pregnancies without OI, highest among which is non-vertex presentation. These data suggest that prenatal studies to identify the type of OI, for determination of mode of delivery alone, may not be of limited value because the major reason to alter mode of delivery is related to obstetric rather than genetic issues.

Reference: Proceedings of the 7th International Conference on Osteogenesis Imperfecta. Montreal, Canada, 1999.