It is now recognized that the absence of one umbilical artery is an important index for detecting many congenital malformations at birth. Single umbilical artery (SUA), one of the most common malformations in man, can be found by the careful inspection of the cut surface of the umbilical cord and is frequently associated with other congenital malformations (Fig. 1). Since Benirschke and Brown (1955) emphasized that SUA was associated with an increased incidence of congenital anomalies, many publications concerning the incidence, the pathogenesis, and the clinical significance of SUA (Benirschke and Bourne, 1960; Gömöri and Koller, 1964; Froehlich and Fujikura, 1966; Papadatos and Paschos, 1965; Peckham and Yerushalmy, 1965; Solnitzky, 1967; Cederqvist, 1970; Soma and Yoshida, 1970; Molz, 1971; Bryan and Kohler, 1974 and Phillippe, 1974).

The reported incidence of SUA, as well as the frequency and type of associated congenital malformations, varies widely. As Benirschke pointed out, there remain some question-
able points concerning the difference of the frequency of SUA in the numerous case reports, the difference of pathogenesis of SUA (whether resulting from primary aplasia or atrophy), the relationship between SUA and chromosome errors, the prognosis of infants with SUA; and follow-up studies of surviving infants (Benirschke and Driscoll, 1967; Benirschke, 1973). Therefore, the clinical and pathologic significance of SUA are reevaluated in this review.

I. The Frequency of SUA

Recent data since 1968, concerning the incidence of SUA, are summarized in Table 1. From these data it is apparent that there is variation in the incidence of SUA and that it occurs with a frequency of slightly less than 1%. It has been found that SUA is more frequent in whites than in blacks and orientals (Peckham and Yerushalmy, 1965; Johnsonbaugh, 1973), and in the present series it also appears that the incidence of SUA in Japanese is lower than in whites (Itoh et al., 1976; Soma and Yoshida, 1977). In addition, our recent data have been tended to be lower than previously ascertained incidence figures (Soma et al., 1963). It is suggested that such discrepancies in frequency of SUA reported by many authors might have arisen because of different methods of examination of the cords by various investigators. According to Kristoffersen (1969), the incidence of SUA is found to be significantly higher on macroscopic or microscopic examinations after fixation than that found by routine inspection of fresh cords at birth. It is concluded that the best way to discover all cases of SUA is to examine all cords macroscopically or microscopically after fixation in formalin or glacial acetic acid.

It has been generally stated that SUA is more frequently associated with twins: According to an analysis of 250 consecutive twin placentas, 18 infants exhibited SUA — a frequency of 3.6% (Benirschke, 1965). From our data of 108 twin placentas, SUA appeared with as high an incidence as 5.6% compared to 0.52% in the singleton placenta (Matayoshi, 1977).

II. Sex and Birthweight

Birth weight and duration of gestation seem to be determined at least in part by fetal factors. Diminished birth weight is a more important feature of SUA than is the duration of gestation (Solnitzky, 1967). It has also been noted that the incidence of SUA progressively decreases with increments of birth weight (Froehlich and Fujikura, 1966), and the highest incidence of SUA occurs in infants weighing 3.6 lb or less (Peckham and Yerushalmy, 1965). On the other hand, in our series of 34 SUA cases, the incidence of SUA in infants exceeding birth weights of 2500 g was 64.7% and correlated with gestational age (35 weeks and over) (Soma and Yoshida, 1977). However, the incidence of SUA in stillborn infants weighin 2100 g or less was consistently high. As reported by