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# Do well infants born with an isolated single umbilical artery need investigation?


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You are the paediatric house officer performing discharge examinations on the postnatal ward. You are informed of this term neonate whose umbilical cord was noted to have a single umbilical artery (SUA) at delivery. He is otherwise well. You cannot detect any abnormalities on physical examination. Historically, SUA has been said to be associated with congenital malformations of different organ systems. You wish to appraise the evidence whether or not this infant needs investigations to detect associated malformations.

### Structured clinical question

In a term neonate with no other obvious congenital malformations [patient] does the presence of a single umbilical artery [risk factor] necessitate further investigation [intervention] to exclude associated malformations [outcome]?

### Search strategy and outcome

Primary source: Medline via Pubmed using keyword ''umbilical artery''. A total of 477 individual articles were found. This was limited to 152 articles by selecting those in English language and human studies relating to neonates (birth–1 month). The search was verified by using (MeSH) subject heading: ''umbilical artery'' + subheading: abnormalities. Individual abstracts were read. A systematic review with meta-analysis of the relevant studies which matched our structured clinical question was found. The meta-analysis and original articles of seven relevant included studies were appraised.

Secondary sources: Cochrane database and Best Bets. No further papers were identified.

See table 1.

### Commentary

Single umbilical artery has long been recognised as a soft marker for chromosomal abnormalities and congenital malformations. Autopsy series from aborted and still born fetuses report a high incidence of associated malformations. It is therefore conceivable that if SUA is detected in a neonate with obvious physical abnormalities, full investigatory work up to detect occult malformations of various organ systems has to be undertaken. Nevertheless, in many cases SUA can be an isolated feature. It is unclear if apparently asymptomatic infants with SUA need to be investigated.

The meta-analysis cited[1] was a review of 37 studies published over the past 40 years. Eleven of the 37 studies were performed on specimens obtained from autopsy studies of abortusus and stillborn babies. These were not relevant to our question. In the remaining 26 studies, the diagnosis of SUA was made by clinical examination of the placenta or umbilical cord after delivery and thus satisfied our initial criteria. But in only seven of these was there data for asymptomatic isolated SUA. Overall, a mean of 16.2% of infants with isolated SUA had a renal anomaly (median 5.3%). In half these cases (8%) these malformations were severe and persistent on follow up. The most frequent major renal anomaly was vesico-ureteric reflux, grade 2 or greater, in 2.9% of the total population.

In the study by Bourke and colleagues,[2] infants with isolated SUA had a screening ultrasound scan. Those with abnormal scans underwent a micturating cysturethrogram and urine cultures. Vesico-ureteric reflux (VUR) was documented in 4.5% of these infants. It is interesting to note that three of the five infants with VUR developed urinary tract infections (UTI) within the first five months of life.

The incidence of occult renal anomalies in the general paediatric population is about 2.5%;[9] the prevalence of VUR in healthy individuals is unclear. Ransley,[10] in a compilation of several publications, reports a rate of 1.3%.From the currently available evidence it seems that the incidence of silent renal abnormalities in infants with isolated SUA is at least threefold higher for severe malformations and sixfold higher for any renal malformation compared to the general paediatric population. VUR is probably up to three times commoner in these infants. A screening renal ultrasound scan may be useful in detecting occult structural malformations of the urinary tract. However, its positive predictive value in suggesting VUR was low; it was reported as 32.5% in a recent study.[11] As VUR and UTI are believed to be forerunners of reflux nephropathy, it seems prudent to investigate infants born with an isolated SUA by means of a micturating cystourethrogram (MCUG) and maintain a low threshold to diagnose and treat urinary tract infections.

### CLINICAL BOTTOM LINE

- There is an increased proportion of significant occult renal malformations in asymptomatic infants born with an isolated single umbilical artery (8% total population).
- A significant proportion of such infants may have vesico-ureteric reflux (grade 2 or worse).
- Screening renal ultrasonography and micturating cystourethrography are useful investigations to detect associated renal anomalies in these cases.
- There is a lack of data regarding malformations of other organ systems in infants with asymptomatic isolated SUA.


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**Table 1** Do well infants born with an isolated single umbilical artery need investigation?

| Citation | Study group | Level of evidence | Outcome | Key results | Comments |
|---|---|---|---|---|---|
| Thummala *et al* (1998) | 204 infants with isolated single umbilical artery from 7 studies where in infants with isolated single SUA were investigated for occult renal malformations | Meta-analysis of case series (level 3a) | Detection of associated malformations | 33/204 infants had occult renal malformations<br>Mean 16.2%, (95% CI for mean 7.7% to 25.6%; Median 5.3%; (range 0% to 33%)<br>15/204 had major anomalies (7.4%)<br>The most frequent renal anomaly of significance was vesico -ureteric reflux | None of the case series included had controls<br>Only articles in English language were included in the meta-analysis<br>There is no data on other organ system malformations |
| Bourke *et al* (1993) | Prospective case series of 112 infants with isolated SUA from 35 000 deliveries. Case detection was by clinical examination of the placenta<br>All cases underwent screening renal ultrasonography. Those with abnormalities were further investigated with a micurating cystourethrogram (MCUG) and had monthly urine cultures for 6 months | Case series (level 4) | Urinary tract anomalies detected on ultrasonogram | 19/112 had some form of renal anomaly (16.9%). In 8 of them the abnormalities were significant (7.1%). 5/8 had VUR. 3/8 infants had UTI within the first 5 months of age | Included in Thummala paper<br>Does not specify if deliveries were consecutive<br>No control group |
| Leung and Robson (1989) | Case series of 159 infants detected to have SUA from records of 56 919 deliveries during a 20 year period. 27 of these 159 infants who had an isolated SUA underwent renal imaging. | Case series (level 4) | Urinary tract anomalies detected on ultra sonogram or intravenous pyelography (IVP) | 5/27 had abnormal renal imaging (18.5%)<br>One each had multicystic kidneys, hypoplastic kidneys, horse shoe kidneys, hydronephrosis and bifid ureter | Included in Thummala paper<br>Retrospective review<br>Screening tool not the same for all cases<br>No control group |
| Feingold *et al* (1964) | Prospective case series. 32 infants detected to have SUA among 6080 deliveries. Three infants died in the neonatal period. IVP was performed on 24 of the 29 survivors without overt renal malformations | Case series (level 4) | Urinary tract anomalies detected on IVP | 8/24 infants; ( 33.3%) had renal malformation<br>In half of them malformations were severe<br>These included massive reflux with hydronephrosis,absent kidney, horse shoe kidney and severe bladder neck obstruction | Included in Thummala paper<br>Not all cases were investigated<br>No control group |
| Vlietinck *et al* (1972) | Prospective case series without controls. 29 infants were detected to have SUA among 2572 deliveries. 4 were stillborn and 2 died in the neonatal period. 19 of the 23 infants who had an isolated SUA were investigated | Case series (level 4) | Urinary tract anomalies detected on IVP | 1/19 infants (5.3%) had an abnormality—complete duplication of the left renal pelvis | Included in Thummala paper<br>Not all cases were investigated<br>No control group |
| Harris and Van Leeuwen (1968) | Prospective case series without controls. 11 infants detected to have isolated SUA among 2800 consecutive deliveries | Case series (level 4) | Urinary tract anomalies detected on IVP | None of the infants had renal malformations (0/11) | Included in Thummala paper<br>Small sample size<br>No control group |
| VanLeeuwen *et al* (1967) | Prospective case series without controls. 4 infants were detected to have isolated SUA among 2000 consecutive deliveries | Case series (level 4) | Urinary tract anomalies detected on IVP | None of the infants had renal malformations (0/4) | Included in Thummala paper<br>Small sample size<br>No control group |
| Johnsonbaugh (1973) | Prospective case series. 8 infants of 1152 deliveries had isolated SUA. Only 5/8 infants were investigated | Case series (level 4) | Detection occult renal anomalies by IVP, transumbilical artery aortography to detect aortic malformations and chromosomal analysis | None of the 5 investigated infants had renal, aortic malformations or any chromosomal abnormality | Included in Thummala paper<br>Not all cases were investigated<br>Small sample size<br>No control group |


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