

International PPB Registry and Literature Cases of PPB Associated with Cystic Nephroma and other Cystic Renal Lesions

16 Registry and 6 literature cases of PPB have been associated with 22 cases of cystic nephroma and 8 other non-biopsied cystic renal lesions.

Cystic nephroma (or other renal lesions) have occurred in the PPB patients or young relatives: siblings, parents, grandparents, cousins, aunts/uncles, great aunt. One relative (mother of a PPB patient) had Wilms tumor.

PPB Associated with Cystic Nephroma

The PPB may be Type I, Type Ir, II, or III. The age at diagnosis for PPB has been typical for the PPB Type. The course of PPBs in these cases is typical for the PPB Type.

Cystic Nephroma Associated with PPB

The renal lesions have been diagnosed under age 48 months. The renal lesions have had a pathologically benign course, but in two cases have progressed to extensive lesions: one child has had 1.5 kidneys removed; another child had 2 kidneys removed and has undergone renal transplantation.

PPB CN Association Data

For cases on which the Registry has specific data:

PPB cases associated with CN are unilateral 14, bilateral 5.

CN cases associated with PPB are unilateral 27, bilateral 4.

The CN is in the PPB patient in 10 cases; the CN is in a relative of the PPB patient in 6 cases.

In 5 cases, a PPB patient had CN and hamartomatous ("juvenile") small bowel polyps.

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Registry Publications:

2008 Priest [in press]; 2006 Boman; 1996 Priest

Literature cases, see Literature Sources (pathology material on some of these published cases has been reviewed by Registry pathologists, assigned Registry numbers and included in Registry tallies):

2006 Yuksel; 2006 Bouron-Dal Soglio; 2005 Rypens; 2005 Bal; 2000 Ishida; 1999 Lallier; 1995 Cottingham; 1993 Lopez-Andreu; 1993 Delahunt; 1989 Joshi; 1987 Graham; 1984 Holland-Moritz; 1979 Abt; 1964 Williams