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Evolutive profile of pauci-symptomatic forms of Mc Cune Albright syndrome

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In young girls, the occurrence of secretory ovarian cysts may be the first manifestation of Mc Cune Albright Syndrome.

We reported the evolutive profile of 8 young patients with secretory ovarian cyst and peripheral precocious puberty.

5 of the 8 girls present metrorrhagia at the diagnostic. 2 girls had café-au-lait spots (cases 4, 7). No patient had bone lesions detected on the holoskeleton.

On the first episode of cyst, the mean age was 3.8 years (range 2.5 to 7.25 years).

The average diameter of the ovarian cyst was 38.5 mm (range 25 to 80 mm).

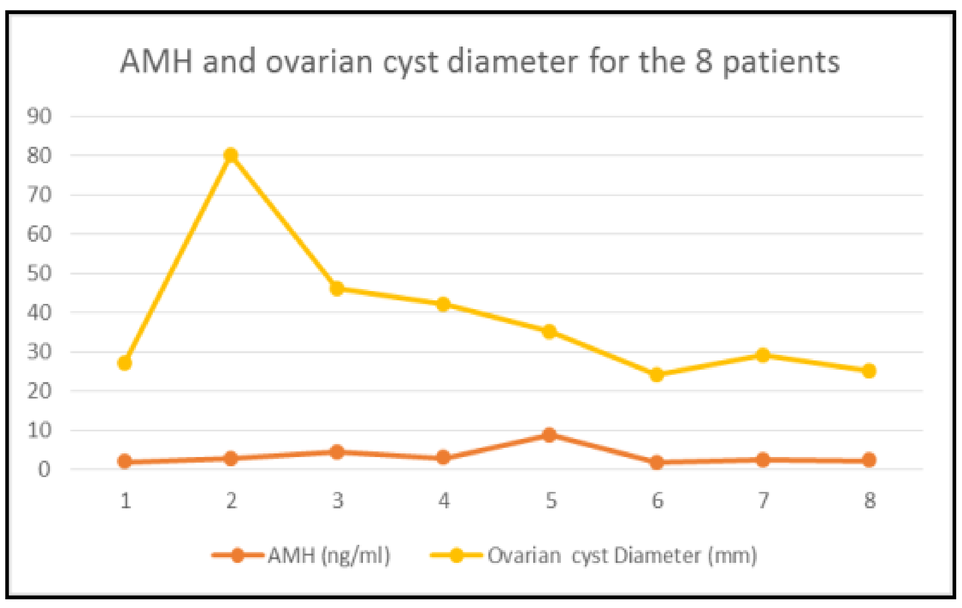
The mean estradiol level was 32.5 pg/ml (range 3 to 160). The mean AMH level was 3.35 ng/ml (range 1.9 to 8.7). The rates of E2 and AMH level were not correlated to the diameter of the cyst.

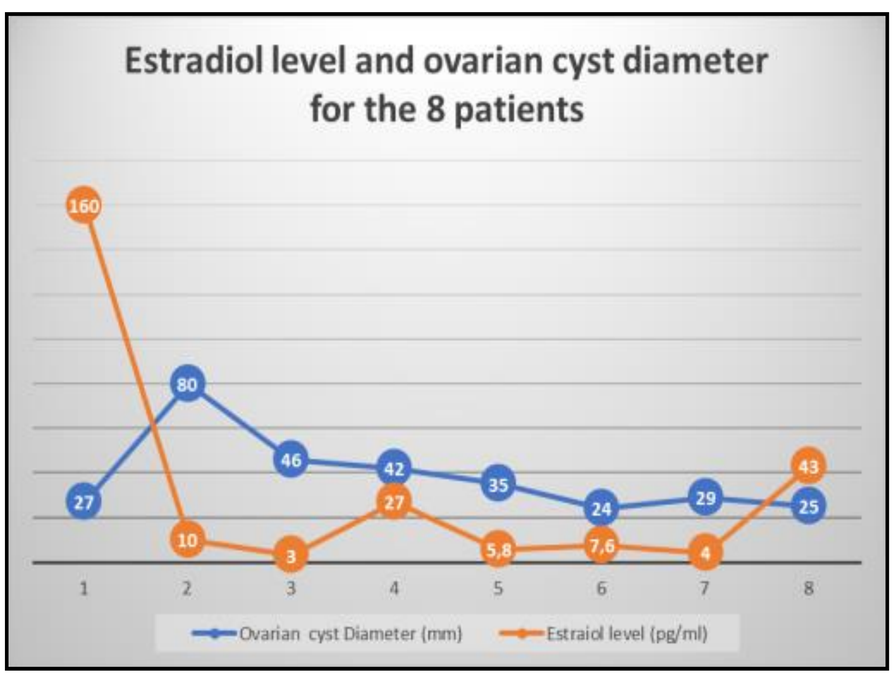
No patient had detected GSα protein mutation by peripheral blood analysis.

5 patients underwent cystectomy (cases 1,2, 3 and 4). GSα protein mutation was positive on the follicular fluid for 4 cases and negative for 1 case. The cyst spontaneously regressed in the 3 other cases (cases 5, 6, 7).

The mean follow up is 6,6 years. The recurrence of the cyst was noted once (case 7 at 4 yrs), twice (case 4 at 7 and 10 yrs) and 6 times (case 1 at 3, 4, 6, 7, 9 and 11 yrs). This girl with 6 recurrences (case 1) was followed until the age of 27 years, and she had no other recurrence of cyst after 11 years.

Patients	1	2	3	4	5	6	7	8
Age at 1st episode (yrs) Tanner	2 yrs 5m	7 P2B2	1yrs 8 m P1B2	4 P1B2	3yrs 9m P2B2	5 P1B3	3 P2B2	3yrs 5m P2B2
Metrorrhagia	+	+	0	+	+	0	0	+
Diameter of the cyst (mm)	27	80	46	42	35	24	29	25
Estradiol (pg/ml) LH/FSH (U/L) AMH (ng/ml)	160 1.9	10 0.2/0.3 2.7	3 0.2/0.3 4.4	27 0.2/03 2.8	5.8 0.6/0.2 8.7	7.6 0.3/0.3 1.7	4 0.2/0.3 2.4	43 0.1/0.1 2.2
Evolution of initial cyst Regression (R) Cystectomy (C)	C	C	C	C	R	R	R	C
Positive GSa protein mutation in follicle cyst fluid	+	+	+	+				-
Number of cyst recurrence	6	0	0	2	0	0	1	0
Age at Recurrence (yrs)	3;4;6;8;7;2;9;11			7;10			4	
Follow up (years)	25	5	8	7	4	1	2	3





Références:

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Conclusion: Mc Cune Albright syndrome is a sporadic disease with unpredictable evolution. We report here cases of 8 young girls with secretory ovarian cyst. There is no predictive factor of recurrence at the initial diagnosis (AMH, Estradiol, cyst diameter).

We conclue that follow-up is essential after a first secretory ovarian cyst before puberty.