P51. Pregnancy stimulates cerebellar hemangioblastoma growth in von Hippel-Lindau disease

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Table 1
VHL complications during pregnancy.

Patient	Birth child (year)	VHL known before pregnancy (yes/no)	Lesion	Intervention
1	1984 Fetal death	No	Bilateral pheochromocytoma (revealed after delivery)	Bilateral adrenalectomy (6 months after delivery)
2	1999	No	Bilateral pheochromocytoma	Caesarean section combined with adrenalectomy (28 weeks gestation)
3	2008	Yes	Cerebellar hemangioblastoma and hydrocephalus Retinal angioma	Caesarean section combined with craniotomy (35 weeks gestation) Eye laser treatment (22 weeks gestation)
4	2007	No	Cerebellar hemangioblastoma and hydrocephalus	Craniotomy (1 week after delivery)
5	2009	Yes	Myelum hemangioblastoma	Resection hemangioblastoma (6 months after delivery)
6	2004	Yes	Renal cell carcinoma Ablatio retinae	Partial nephrectomy right sided (20 weeks gestation) Victrectomy (10 weeks gestation)
7	1979	No	Ablatio retinae – papillary angioma (known before pregnancy)	–
8	2000	Yes	Ablatio retinae – angioma (known before pregnancy)	–

other group when this I defined at 6 week weight expressed as a percentage of their birthweight ($p = 0.001$). 31% of the babies across all the groups showed catch up growth (a gain in z score for weight greater than 0.67 z score) in the first 6 weeks of life. Babies of mothers with PE have a higher head circumference to birth weight ratio (brain sparing) than babies from GH or EH groups ($p = 0.007$).

Conclusions: Babies born to mothers with PE with or without underlying EH were smaller at birth but showed evidence of brain sparing. 30% of babies born after hypertensive pregnancy are at increased risk of obesity in childhood and a potential risk for disease in adulthood by risk of catch up growth.

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Objectives: von Hippel-Lindau (VHL) disease is an autosomal dominant disorder associated with the development of multiple benign and malignant tumors throughout life – hemangioblastomas in the central nervous system (CNS), cysts and tumors in the visceral organs, pheochromocytomas in the adrenal glands. Case reports make note of complicated pregnancy and/or delivery by CNS hemangioblastomas and pheochromocytomas. Maternal pheochromocytomas induced hypertension can be mistaken for a more common pregnancy related hypertensive disorder. One retrospective study, based on questionnaires described a VHL disease related maternal morbidity rate during pregnancy of 5%. Progression of VHL lesions during pregnancy may be attributed to altered hemodynamic and hormonal states, although evidence is lacking. Aim of our study was to examine the course of pregnancy in VHL disease.

Materials and methods: We studied VHL patients with at least one delivery with follow-up in VHL-expertise centers in the Netherlands. The medical status of all patients was reviewed to quantify primary pregnancy outcome and maternal and VHL related complications. The effect of pregnancy on individual VHL lesions was determined. If available, imaging reports (MRI cerebellum and myelum/CT and ultrasound abdomen/fundoscopy/serum or urine metanephrine levels) were used to assess a lesion progression score before and after pregnancy in different organs. These scores were analyzed by the Friedman test (p 0.05).

Results: Twenty-nine patients were studied, with 48 pregnancies and 49 newborns. Fetal mortality rate was 2% ($n = 1$) caused by maternal pheochromocytoma induced hypertension. Maternal VHL related complications occurred in 17% ($n = 8$) of all pregnancies (Table 1), in half of the patients their VHL was known before pregnancy. In four of these a life threatening situation emerged during their first pregnancy: pheochromocytoma ($n = 2$) and hydrocephalus due to a cerebellar hemangioblastoma ($n = 2$). In both pheochromocytomas VHL disease was not yet known before pregnancy. Maternal pregnancy-related hypertensive disorders occurred in 12.5% ($n = 6$). The progression score of hemangioblastomas in the cerebellum was significantly different in one MRI before and two after pregnancy ($p = 0.049$) ($n = 12$), illustrating an initial increase and in the second MRI after delivery a decrease in progression. In other organs no significant progression of lesions was found.

Conclusion: Our results point to a relevant clinical conclusion: progression of VHL cerebellar hemangioblastomas during pregnancy was shown, and uncontrolled hypertension in pregnancy can be caused by a pheochromocytoma. So intensified screening of VHL patients in a specialized medical centre is recommended during preconception care and during pregnancy.

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