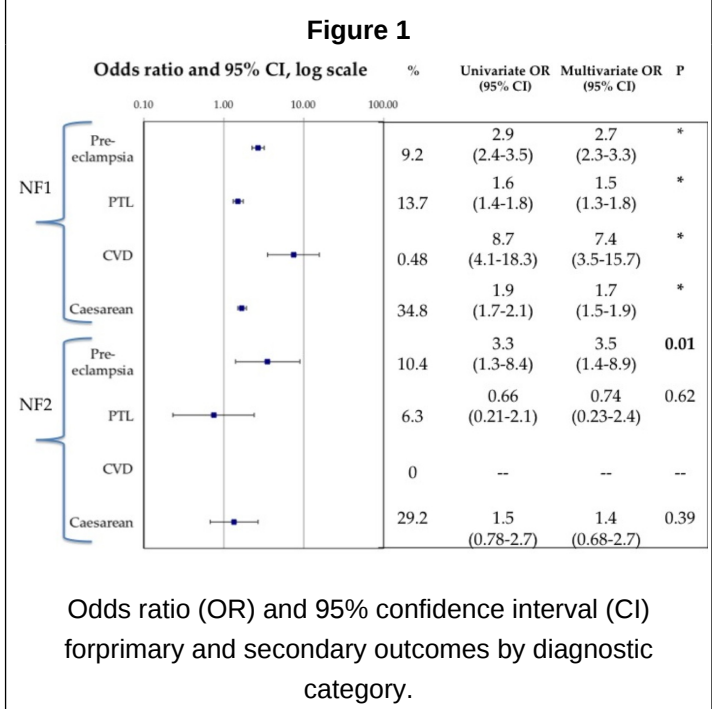




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PTL = preterm labor
CVD = cerebrovascular disease
* P < 0.0001

Conclusions

We successfully demonstrated the use of a large database to study pregnancy outcomes in a rare disease. NF1 was associated with a higher risk of vascular complications of pregnancy, possibly due to an underlying vasculopathy that has been reported in NF1 patients. Pre-eclampsia is a complex disorder of endothelial dysfunction associated with earlier onset of hypertension. Further research is needed to clarify the mechanisms of obstetric and vascular risk in NF.

	NF1	NF2	Control
Characteristic	N=1450	N=48	N=18,716,102
Age			
Mean (SD)	26.4 (5.8)	26.6 (5.8)	27.1 (6.2)
Young (<20 years), %	10.6	<10 ¹	11.9
Old (>40 years), %	1.9	<10	2.2
Race, %			
White	40.9	50.0	38.8
Black	17.2	<10	10.1
Hispanic	8.1	<10	14.3
Other	3.4	<10	5.8
Missing	30.4	20.8	30.9
Income quartile, %			
1 (least wealthy)	31.0	<10	23.7
2	27.3	<10	22.4
3	19.9	22.9	20.9
4 (most wealthy)	17.8	39.6	30.4
Missing	4.0	<10	2.6
Comorbidities, %			
Diabetes ²	0.7	0	1.0
Hypertension	8.4	<10	4.0
Renal disease	0.9	0	0.3
Multiple gestation	1.6	0	1.4
Hospital setting³, %			
Rural	13.4	<10	11.6
Urban non-teaching	33.2	27.1	46.7
Urban teaching	53.2	68.8	41.5
Hospital size³, %			
Small	8.6	<10	11.6
Medium	24.9	29.2	28.3
Large	66.3	68.8	60.0

1. Exact number cannot be reported when cell size < 10.
2. Non-gestational.
3. Less than 1% missing data.

Results

NF1 was associated with higher odds of pre-eclampsia, preterm labor, cerebrovascular disease, and Caesarean delivery. For NF2, only the risk of pre-eclampsia was elevated, and there were no events of cerebrovascular disease.

Whether patients with neurofibromatosis (NF) are at higher risk for adverse pregnancy outcomes is an important but understudied clinical question. Previous studies have been limited by small sample size and lack of adequate follow-up. Due to the higher prevalence of tumors and vascular disorders in NF, we hypothesized that adverse outcomes of pregnancy might occur at a higher rate in these patients.

Methods

Data source: The Nationwide Inpatient Sample (NIS) is a large administrative database representing a 20% stratified sample of all nonfederal US hospital discharges (approximately 8 million per year).

Study design: Using the NIS, we performed a retrospective cohort study of hospitalizations of pregnant patients with NF1 and NF2.

Case selection: All hospitalizations of patients with an associated pregnancy-related diagnosis were included in the control population. Patients with NF1 and NF2 were identified as separate cohorts by their relevant ICD-9 codes. Outcomes of interest included pre-eclampsia, preterm labor, cerebrovascular disease, and Caesarean delivery.

Data analysis: We constructed a logistic regression model, adjusting for extremes of age, diabetes, hypertension, renal disease, multiple gestation, prior Caesarean delivery, demographics, and hospital characteristics. Outcomes and covariates were defined using NIS variables and ICD-9-CM diagnosis and procedure codes.