**Table I-1. Overview of Maternal PKU Collaborative Study (MPKUCS) papers**

| **Author, Year** | **Title** | **Study Date** | **Countries** | **# of Women** | **# of Pregnancies** | **Cognitive Outcomes Reported** |
| --- | --- | --- | --- | --- | --- | --- |
| **Studies Reporting Cognitive Outcomes** |
| Rouse et al., 20041  | Effect of High Maternal Blood Phenylalanine on Offspring Congenital Anomalies and Developmental Outcome at Ages 4 and 6 Years: The Importance of Strict Dietary Control Preconception and Throughout Pregnancy | NR | USCanada | 526100 matched controls | 576413 live births | * McCarthy at 4 yrs
* WISC-R at 6 yrs
 |
| Guttler et al., 20032 | Impact of the Phenylalanine Hydroxylase Gene on Maternal Phenylketonuria Outcome | NR | USCanadaGermany | 196 fully evaluated;236 genotyped | 308 pregnancies;253 live births;196 offspring fully evaluated | * WISC-R at 6-7 yrs or later
 |
| Levy et al., 20033 | Pregnancy Experiences in the Woman With Mild Hyperphenylalaninemia | NR | USCanadaGermany | 48 | 58 | * WISC-R at 2 years
* McCarthy Scales of Children’s Abilities
* Bayley Scales of Infant Development
 |
| Waisbren et al., 20034 | Cognitive and Behavioral Development in Maternal Phenylketonuria Offspring | NR | USCanadaGermany | NR | NR;228 offspring;70 controls | * WISC-R at 7 years
* Test of Language Development, 2nd edition
* Peabody Individual Achievement Test-Revised
* Stroop Interference
* Visual Motor Integration
* Child Behavior Checklist
 |
| Koch et al., 20035 | The Maternal Phenylketonuria International Study: 1984-2002 | 1984-2002 | USCanadaGermany | 382 | 572 | * McCarthy at 4yrs
* WISC-R at 7 yrs
* WISC-R at 10 yrs
 |
| Widaman et al., 20036 | Relation of Prenatal Phenylalanine Exposure to Infant and Childhood Cognitive Outcomes: Results From the International Maternal PKU Collaborative Study | NR | USCanadaGermany | NR | 572 pregnancies;413 offspring evaluated | * Bayley at 1 year
* McCarthy at 4 years
* TOLD CSLQ at 4 years
* WISC-R at 7 years
 |
| Antshel et al., 20037, 8 | Developmental Timing of Exposure to Elevated Levels of Phenylalanine is Associated with ADHD Symptom Expression | NR | US | NR | 15 MPKU offspring;46 PKU patients;18 controls | * ADHD symptoms
* FSIQ
 |
| Platt et al., 20009 | The International Study of Pregnancy Outcome in Women with Maternal Phenylketonuria: Report of a 12-year study | 1984-1999 | USCanadaGermany | PKU NR;101 controls | 576 pregnancies;414 live births | * Bayley at 2 yrs
* McCarthy at 4-5 yrs
* WISC-R data presented on children who had reached age 7
 |
| Koch et al., 200010 | The international collaborative study of maternal phenylketonuria: status report 1998 | 1984-1998 | USCanada | NR | 572 HPA99 control | * Congenital anomalies
* McCarthy at 4 years
 |
| Waisbren et al., 200011 | Outcome at Age 4 Years in Offspring of Women With Maternal Phenylketonuria: The Maternal PKU Collaborative Study | 1984-2000 | USCanada | 205253 offspring received preschool evaluation | 572 pregnancies;412 offspring | * McCarthy at 3-5 years
* Test of language development at 4 years
* Achenbach at 4 years
* Vineland at 4 years
* Bayley at 2 yrs
 |
| Waisbren, et al., 199812 | Neonatal neurological assessment of offspring in maternal phenylketonuria | NR | US | NR | 56 PKU offspring;45 control offspring | * Dubowitz Neurological Assessment of the Preterm and Full-Term Newborn Infant
* Bayley Mental and Motor Scales at 1 year
* Receptive-Expressive Emergent Language Scales at 1 year
 |
| Hanley et al., 199613 | The North American Maternal Phenylketonuria Collaborative Study, developmental assessment of the offspring: preliminary report | 1984-1994 | USCanada | NR | 134 HPA offspring;58 control offspring | * Bayley at 2 yrs
* McCarthy at 4 yrs
 |
| Cipcic-Schmidt et al., 199614 | German Maternal Phenylketonuria Study | 1989-1994, worked with North American study since 1992 | GermanyAustria | 275 | 43 pregnancies;34 live births | * Bayley tests at 2 yrs
 |
| Koch et al., 199415 | The international collaborative study of maternal phenylketonuria: status report 1994 | 1984-1994 (US)1986-1994 (Canada)1993-1994 (Germany) | USCanadaGermany | NR | 402 pregnancies;99 control | * Bayley Scales and McCarthy at 3.5 to 4.5 yrs
 |
| Koch et al., 199316 | The North American Collaborative Study of Maternal Phenylketonuria: Status Report 1993 | 1984-1992 | USCanada | 379 HPA | 318 HPA;59 control | * Bayley scale at 12-24 mos
* McCarthy Scale between 3.5-5yrs
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ADHD=attention deficit hyperactivity disorder; FSIQ=full scale intelligence quotient; MPKU=maternal phenylketonuria; NR=not reported; PKU=phenylketonuria; TOLD CSLQ=Test of Language Development Spoken Language Component; WISC-R=Wechsler Intelligence Scale for Children-Revised