low phenylalanine content of breast milk, total or partial breastfeeding may be possible with close monitoring of phenylalanine levels (Lawrence and Lawrence, 2011).

When treatment for PKU was first instituted, it was believed that phenylalanine withdrawal during only the first 3 years of age would suffice to avoid cognitive impairment and other deleterious manifestations of PKU. However, most clinicians now agree that to achieve optimal metabolic control and outcome, a restricted phenylalanine diet, including medical foods and low-protein products, most likely will be medically required for virtually all individuals with classic PKU for their entire lives (Soltanizadeh and Mirmoghtadaie, 2014). Such lifetime reduction of phenylalanine intake is necessary to prevent neuropsychological and cognitive deficits because even mild hyperphenylalaninemia (20 mg/dl) would produce such effects. To evaluate the effectiveness of dietary treatment, frequent monitoring of blood phenylalanine and tyrosine levels is necessary.

Phenylalanine levels greater than 6 mg/dl in mothers with PKU affect the normal embryologic development of the fetus, including cognitive impairment, cardiac defects, and LBW. It is recommended that phenylalanine levels below 6 mg/dl be achieved at least 3 months before conception in women with PKU (Koch, Trefz, and Waisbren, 2010).

Prognosis

Although many individuals with treated PKU manifest no cognitive and behavioral deficits, many comparisons of individuals with PKU with control participants show lower performance on IQ tests, with larger differences in other cognitive domains; however, their performance is still in the average range. Evidence for differences in behavioral adjustment is inconsistent despite anecdotal reports suggesting greater risk for internalizing psychopathology and attention disorders. In addition, insufficient data are available on the effects of phenylalanine restriction over many decades of life (Kaye, Committee on Genetics, Accurso, et al, 2006). Recent data suggest that treatment with tetrahydrobiopterin in addition to the phenylalanine-restricted diet may be beneficial to PKU patients (Blau, van Spronsen, and Levy, 2010). Total bone mineral density is considerably lower in children who are on a low-