

Beaujon Extralight

Beaujon Light

Beaujon Regular

Beaujon Medium

Beaujon Bold

Beaujon Extrabold

Beaujon Extralight Italic

Beaujon Light Italic

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Tetrahydrobiopterin (BH₄, THB), also known as sapropterin (INN), is a cofactor of the three aromatic amino acid hydroxylase enzymes, used in the degradation of amino acid phenylalanine and in the biosynthesis of the neurotransmitters serotonin (5-hydroxytryptamine, 5-HT), melatonin, dopamine, norepinephrine (noradrenaline), epinephrine (adrenaline), and is a cofactor for the production of nitric oxide (NO) by the nitric oxide synthases. Chemically, its structure is that of a (dihydropteridine reductase) reduced pteridine derivative (quinonoid dihydrobiopterin). Tetrahydrobiopterin is available as a tablet for oral administration in the form of sapropterin dihydrochloride (BH₄*2HCL). It was approved for use in the United States as a tablet in December 2007 and as a powder in December 2013. It was approved for use in the European Union in December 2008, Canada in April 2010, and Japan in July 2008. It is sold under the brand names Kuvan and Biopoten. The typical cost of treating a patient with Kuvan is US\$100,000 per year. BioMarin holds the patent for Kuvan until at least 2024, but Par Pharmaceutical has a right to produce a generic version by 2020. Sapropterin is indicated in tetrahydrobiopterin deficiency caused by GTP cyclohydrolase I (GTPCH) deficiency, or 6-pyruvoyltetrahydropterin synthase (PTPS) deficiency. Also, BH₄*2HCL is FDA approved for use in phenylketonuria (PKU), along with dietary measures. However, most people with PKU have little or no benefit from BH₄*2HCL. The most common adverse effects, observed in more than 10% of people, include headache and a running or obstructed nose. Diarrhea and vomiting are also relatively common, seen in at least 1% of people. No interaction studies have been conducted. Because of its mechanism, tetrahydrobiopterin might interact with dihydrofolate reductase inhibitors like methotrexate and trimethoprim, and NO-enhancing drugs like nitroglycerin, molsidomine, minoxidil, and PDE5 inhibitors. Combination of tetrahydrobiopterin with levodopa can lead to increased excitability. Tetrahydrobiopterin has multiple roles in human biochemistry. The major one is to convert amino acids such as phenylalanine, tyrosine, and tryptophan to precursors of dopamine and serotonin, major monoamine neurotransmitters. It works as a cofactor, being required for an enzyme's activity as a catalyst, mainly hydroxylases. Tetrahydrobiopterin is a cofactor for

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