

## **Supplementary file S2 - Search strategy**

### **Databases**

Three databases were searched:

1. Pubmed
2. Cochrane
3. Embase (Ovid)

A combination was used of:

- Mesh terms (Medical Subject Headings) from Pubmed and Cochrane or Emtree terms from Embase
- Text-word search in title, abstract and author keywords.

### **Entry terms**

1. "phenylketonurias"[Mesh]
2. "phenylalanine hydroxylase/deficiency"[Mesh]
3. hyperphenylalaninaemia OR hyperphenylalaninemia
4. phenylalanine hydroxylase deficiency OR PAH deficiency
5. phenylketonuri\*
6. pku
7. 1 or 2 or 3 or 4 or 5 or 6
8. "sapropterin" [Supplementary Concept]
9. tetrahydrobiopterin
10. bh4 or bh 4
11. sapropterin
12. kuvan
13. 8 or 9 or 10 or 11 or 12
21. 7 and 13

For Embase the following publications types were excluded: books or chapter or conference abstract or editorial or note or tombstone

### Syntax of Mesh/Emtree terms per database

Entry nr	Pubmed	Cochrane	Embase (OVID)
1	"phenylketonurias"[Mesh]	MeSH descriptor: [phenylketonurias] explode all trees	exp phenylketonuria/ exp hyperphenylalaninemia/
2	"phenylalanine hydroxylase/deficiency"[Mesh]	MeSH descriptor: Phenylalanine Hydroxylase] explode all trees and with qualifier(s): [deficiency – DF]	exp phenylalanine 4 monooxygenase/ and deficiency.ti.ab.kw
8	"sapropterin" [Supplementary Concept]	/	exp tetrahydrobiopterin/ exp sapropterin/

### Syntax of title, abstract and author keyword per database

Entry nr	Pubmed	Cochrane	Embase (OVID)
3-6, 14-23	[tiab]	:ti,ab,kw	.ti,ab,kw.