

Texas Prior Authorization Program Clinical Criteria

Drug/Drug Class

Urea Cycle Disorder Agents

This criteria was recommended for review by an MCO to ensure appropriate and safe utilization

Clinical Criteria Information Included in this Document

Agents for the treatment of Urea Cycle Disorders

- **Drugs requiring clinical prior authorization:** the list of drugs requiring prior authorization for this clinical criteria
- **Clinical prior authorization criteria logic:** a description of how the prior authorization request will be evaluated against the clinical criteria rules
- **Logic diagram:** a visual depiction of the clinical criteria logic
- **Supporting tables:** a collection of information associated with the steps within the criteria (diagnosis codes, procedure codes, and therapy codes)
- **References:** clinical publications and sources relevant to this clinical criteria

Note: Click the hyperlink to navigate directly to that section.

Revision Notes

Annual review by staff

Added GCN for carglumic acid (20522)

Updated references



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Drugs Requiring Clinical Prior Authorization

The listed GCNS may not be an indication of TX Medicaid Formulary coverage. To learn the current formulary coverage, visit TxVendorDrug.com/formulary/formulary-search.

Label Name	GCN
BUPHENYL 500 MG TABLET	43371
BUPHENYL POWDER	43370
CARBAGLU 200 MG DISPER TABLET	20522
CARGLUMIC ACID 200 MG TAB SUSP	20522
OLPRUVA 2 GRAM DOSE ENVELOPE	53445
OLPRUVA 2 GRAM DOSE KIT	53445
OLPRUVA 2 GRAM PACKET	53445
OLPRUVA 3 GRAM DOSE ENVELOPE	53446
OLPRUVA 3 GRAM DOSE KIT	53446
OLPRUVA 3 GRAM PACKET	53446
OLPRUVA 4 GRAM DOSE ENVELOPE	53447
OLPRUVA 4 GRAM DOSE KIT	53447
OLPRUVA 5 GRAM DOSE ENVELOPE	53448
OLPRUVA 5 GRAM DOSE KIT	53448
OLPRUVA 6 GRAM DOSE ENVELOPE	53449
OLPRUVA 6 GRAM DOSE KIT	53449
OLPRUVA 6.67 GM DOSE ENVELOPE	53452
OLPRUVA 6.67 GRAM DOSE KIT	53452
RAVICTI 1.1GRAM/ML LIQUID	34137
SODIUM PHENYLBUTYRATE POWDER	43370
SODIUM PHENYLBUTYRATE 500 MG TAB	43371



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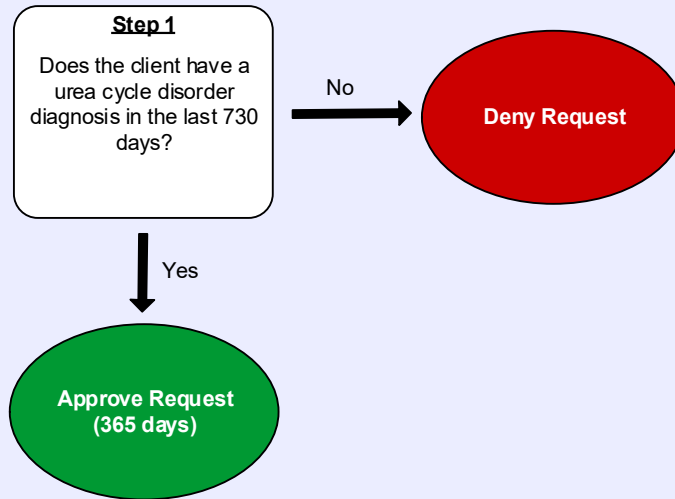
Clinical Criteria Logic

1. Does the client have a **urea cycle disorder diagnosis** in the last 730 days?
 Yes – Approve (365 days)
 No – Deny



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Clinical Criteria Logic Diagram





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Clinical Criteria Supporting Tables

Step 1 (diagnosis of urea cycle disorder) Required quantity: 1 Look back timeframe: 730 days	
ICD-10 Code	Description
E7220	DISORDER OF UREA CYCLE METABOLISM
E7221	ARGININEMIA
E7222	ARGINOSUCCINIC ACIDURIA
E7223	CITRULLINEMIA
E7229	OTHER DISORDERS OF UREA CYCLE METABOLISM
E724	DISORDERS OF ORNITHINE METABOLISM



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Clinical Criteria References

1. Clinical Pharmacology [online database]. Tampa, FL: Elsevier / Gold Standard, Inc. 2021. Available at www.clinicalpharmacology.com. Accessed on September 15, 2021.
2. Drug Facts and Comparisons. eFacts [online]. 2021. Available from Wolters Kluwer Health, Inc. Accessed on September 15, 2021.
3. Ravicti Prescribing Information. Lake Forest, IL. Horizon Therapeutics. November 2019.
4. Buphenyl Prescribing Information. Lake Forest, IL. Horizon Therapeutics. March 2021.
5. Carbaglu Prescribing Information. Lebanon, NJ. Recordati Rare Diseases, Inc. August 2021.
6. Lee B (2020). Urea cycle disorders: Management. In S Hahn (Ed), UpToDate. Accessed September 15, 2021.
7. Olpruva Prescribing Information. Newton, MA. Acer Therapeutics Inc. December 2022.



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Publication History

The Publication History records the publication iterations and revisions to this document. Notes for the *most current revision* are also provided in the **Revision Notes** on the first page of this document.

Publication Date	Notes
04/26/2019	Initial publication and presentation to the DUR Board
01/12/2022	Annual review by staff Updated references
07/07/2023	Added GCNs for Olpruva (53445, 53446, 53447, 53448, 53449, 53452)
02/23/2024	Annual review by staff Added GCN for carglumic acid (20522) Updated references