

# Diagnosis and Treatment of Neurocysticercosis: 2017 Clinical Practice Guidelines by the Infectious Diseases Society of America (IDSA) and the American Society of Tropical Medicine and Hygiene (ASTMH)<sup>1</sup>

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## EXECUTIVE SUMMARY

Guidelines for the clinical management of patients with neurocysticercosis (NCC) were prepared by a panel of the Infectious Diseases Society of America (IDSA) and the American Society of Tropical Medicine and Hygiene (ASTMH). The guidelines are intended for infectious disease specialists, neurologists, neurologist surgeons, internists, pediatricians, and family practitioners.

These guidelines present our approaches to the diagnosis and management of patients with the different forms of neurocysticercosis, including viable parenchymal neurocysticercosis, single enhancing lesions, calcified parenchymal neurocysticercosis, ventricular neurocysticercosis, and subarachnoid neurocysticercosis. Our recommendations are based on the best evidence available. Due to the complex variations in clinical manifestations and the limitations of the literature, many of the recommendations are

based on observational studies, anecdotal data, or expert opinion rather than randomized clinical trials. The approaches we describe are intended to be both applicable and feasible in the United States and Canada (for simplicity, referred to here as North America). The recommendations may not apply for settings where resource constraints may limit their applicability. The executive summary below lists the recommendations for the diagnosis and clinical management of neurocysticercosis. A detailed description of the methods, background, and evidence summaries that support each of the recommendations can be found online in the full text of the guidelines. A criterion for grading evidence is presented in [Figure 1 \[1\]](#). Note that diagnosis and management of patients with neurocysticercosis can be challenging even with expert guidelines. Due to this complexity, clinicians with little experience with this disease should have a low threshold for consultation with an expert in the disease.

## RECOMMENDATIONS FOR DIAGNOSIS AND BASELINE EVALUATION

### I. How should NCC be diagnosed?

#### Recommendations

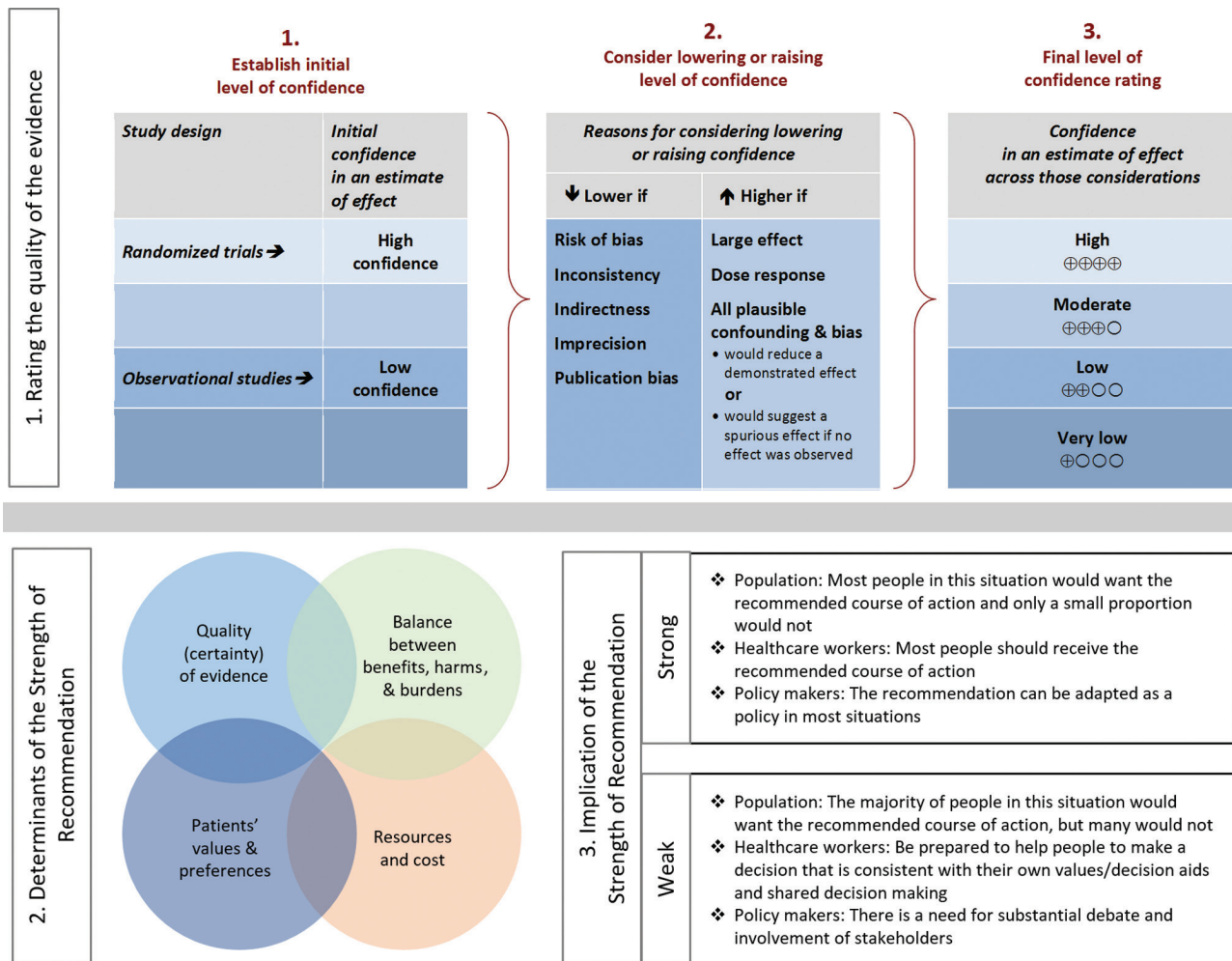
1. While there is a wide range of clinical manifestations of neurocysticercosis, the 2 most common clinical presentations are with seizures and increased intracranial pressure (*not graded*).
2. Initial evaluation should include careful history and physical examination, and neuroimaging studies (*not graded*).
3. We recommend serologic testing with enzyme-linked immunotransfer blot as a confirmatory test in patients with suspected neurocysticercosis (*strong, moderate*). Enzyme-linked immunosorbent assays using crude antigen should be avoided due to poor sensitivity and specificity (*strong, moderate*).

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**Figure 1.** Grading of Recommendations, Assessment, Development and Evaluation (GRADE) framework for grading of evidence.

## II. What imaging studies should be used to classify disease?

### Recommendation

4. We recommend both brain magnetic resonance imaging (MRI) and a noncontrast computed tomography (CT) scan for classifying patients with newly diagnosed neurocysticercosis (*strong, moderate*).

## III. What additional tests should be performed prior to initiation of therapy?

### Recommendations

5. We suggest screening for latent tuberculosis infection in patients likely to require prolonged corticosteroids (*weak, low*).
6. We suggest screening or empiric therapy for *Strongyloides stercoralis* in patients likely to require prolonged corticosteroids (*weak, low*).

7. We recommend that all patients with NCC undergo a fundoscopic examination prior to initiation of anthelmintic therapy (*strong, moderate*).
8. We suggest that patients with NCC who probably acquired NCC in a nonendemic area have their household members screened for tapeworm carriage (*weak, low*). *Remark:* This is a public health issue and can often be addressed by the local health department.

## IV. How should antiparasitic and anti-inflammatory therapy be monitored?

### Recommendations

9. We recommend that patients treated with albendazole for >14 days be monitored for hepatotoxicity and leukopenia (*strong, moderate*).
10. No additional monitoring is needed for patients receiving combination therapy with albendazole and praziquantel beyond that recommended for albendazole monotherapy (*strong, moderate*).

## RECOMMENDATIONS FOR THE TREATMENT OF VIABLE INTRAPARENCHYMAL NEUROCYSTICERCOSIS

### V. What is the role of antiparasitic drugs in viable intraparenchymal neurocysticercosis (VPN)?

#### Recommendations

11. In patients with untreated hydrocephalus or diffuse cerebral edema, we recommend management of elevated intracranial pressure alone and not antiparasitic treatment (*strong, moderate*). *Remarks:* The management of patients with diffuse cerebral edema should be anti-inflammatory therapy such as corticosteroids, whereas hydrocephalus usually requires a surgical approach.
12. In the absence of elevated intracranial pressure, we recommend use of antiparasitic drugs in all patients with VPN (*strong, moderate*).
13. For patients with 1–2 viable parenchymal cysticerci, we recommend albendazole monotherapy for 10–14 days compared to either no antiparasitic therapy (*strong, high*) or combination antiparasitic therapy (*weak, moderate*). *Remarks:* The usual dose of albendazole is 15 mg/kg/day divided into 2 daily doses for 10–14 days with food. We recommend a maximum dose of 1200 mg/day.
14. We recommend albendazole (15 mg/kg/day) combined with praziquantel (50 mg/kg/day) for 10–14 days rather than albendazole monotherapy for patients with >2 viable parenchymal cysticerci (*strong, moderate*).
15. We suggest retreatment with antiparasitic therapy for parenchymal cystic lesions persisting for 6 months after the end of the initial course of therapy (*weak, low*).

### VI. What is the role of anti-inflammatory therapy in management of VPN?

#### Recommendation

16. We recommend adjunctive corticosteroid therapy begun prior to antiparasitic drugs rather than no adjunctive therapy in all patients treated with antiparasitic therapy (*strong, moderate*).

### VII. What is the role of antiepileptic drugs in VPN?

#### Recommendations

17. We recommend antiepileptic drugs in all NCC patients with seizures (*strong, low*).
18. In patients with few seizures prior to antiparasitic therapy, resolution of the cystic lesion on imaging studies, and no seizures for 24 consecutive months, we suggest that tapering off and stopping antiepileptic drugs be considered (*weak, moderate*).
19. In the absence of controlled data, the choice of antiepileptic drugs should be guided by local availability, cost, drug interactions, and potential side effects (*not graded*).

### VIII. What follow-up is recommended after initial antiparasitic therapy for patients with VPN?

#### Recommendation

20. We suggest that MRI be repeated at least every 6 months until resolution of the cystic component (*strong, low*).

## RECOMMENDATION FOR THE TREATMENT OF DEGENERATING INTRAPARENCHYMAL NCC INCLUDING PATIENTS WITH SINGLE ENHANCING LESIONS DUE TO NCC

### IX. What should be the initial approach to the patient with multiple enhancing lesions from NCC?

#### Recommendation

21. We recommend that patients with multiple enhancing lesions and seizures be initially treated with antiepileptic drugs, antiparasitic therapy, and corticosteroids as outlined in the section on viable parenchymal cysticerci (*weak, moderate*).

### X. What is the role of antiepileptic medications in patients with single enhancing lesions (SELs) from cysticercosis with seizures?

#### Recommendations

22. We recommend antiepileptic drugs for all patients with SELs and seizures (*strong, moderate*).
23. In the absence of controlled data, the choice of antiepileptic drugs can be guided by local availability, cost, drug interactions, and potential side effects (*not graded*).
24. In patients who have been seizure free for 6 months, we suggest tapering off and stopping antiepileptic drugs after resolution of the lesion in patients with SELs without risk factors for recurrent seizures (*weak, moderate*). *Remark:* Risk factors for recurrent seizures include residual cystic lesions or calcifications on neuroimaging studies, breakthrough seizures, or >2 seizures.

### XI. What is the role of antiparasitic drugs in patients with SELs?

#### Recommendation

25. We suggest albendazole therapy rather than no antiparasitic therapy for all patients with SELs (*weak, moderate*). *Remarks:* Albendazole (15 mg/kg/day in twice-daily doses up for 1–2 weeks) should be given with meals.

### XII. What is the role of anti-inflammatory therapy in SELs?

#### Recommendation

26. We recommend that patients with SELs treated with antiparasitic drugs also be treated with corticosteroids initiated prior to antiparasitic therapy (*strong, moderate*).

### XIII. How should patients with SELs be followed?

#### Recommendation

27. We suggest that MRI be repeated at least every 6 months until resolution of cystic lesions for patients with SELs (*weak, low*).

## RECOMMENDATIONS FOR THE TREATMENT OF CALCIFIED PARENCHYMAL NEUROCYSTICERCOSIS

### XIV. What should the initial approach be to patients with calcified lesions suggestive of calcified parenchymal neurocysticercosis (CPN)?

#### Recommendation

28. We suggest brain MRI in patients with seizures or hydrocephalus and only calcified parenchymal NCC on CT (*weak, low*).

### XV. What is the role of antiparasitic drugs, antiepileptic drugs, and anti-inflammatory medications in the management of patients with CPN?

#### Recommendations

29. We recommend symptomatic therapy alone instead of antiparasitic drugs in patients with calcified parenchymal lesions (*strong, moderate*).
30. We suggest that corticosteroids not be routinely used in patients with isolated CPN and perilesional edema (*weak, low*).

### XVI. Is there a role for surgical therapy in refractory cases?

#### Recommendation

31. In patients with refractory epilepsy and CPN, we suggest evaluation for surgical removal of seizure foci (*weak, low*).

## RECOMMENDATIONS FOR THE TREATMENT OF INTRAVENTRICULAR NEUROCYSTICERCOSIS

### XVII. How are extraparenchymal cysts best identified?

#### Recommendation

32. We recommend MRI with 3D volumetric sequencing to identify intraventricular and subarachnoid cysticerci in patients with hydrocephalus and suspected NCC (*strong, moderate*).

### XVIII. What is the optimal approach to management of intraventricular neurocysticercosis (IVN) in the lateral and third ventricles?

#### Recommendation

33. When possible, we recommend removal of the cysticerci by minimally invasive neuroendoscopy over other surgical or medical approaches for cysticerci of the lateral and third

ventricles (*strong, moderate*). *Remark:* Most experts recommend that antiparasitic drugs not be used preoperatively, as such treatment could result in disruption of parasite integrity and an inflammatory response that could prevent successful cyst removal.

### XIX. What is the optimal surgical approach to management of IVN in the fourth ventricle?

#### Recommendation

34. In cases in which surgical removal of fourth ventricular cysticerci is possible, we recommend surgical removal rather than medical therapy and/or shunt surgery (*strong, moderate*).

### XX. What is the optimal approach to adherent IVN?

#### Recommendation

35. We suggest shunt surgery for hydrocephalus rather than cyst removal when surgical removal is technically difficult (*weak, low*). *Remark:* Attempted removal of inflamed or adherent ventricular cysticerci is associated with increased risk of complications.

### XXI. Does medical therapy as an adjunct to procedures or as primary therapy have an impact on outcome in treating patients with IVN?

#### Recommendations

36. We recommend corticosteroids to decrease brain edema in the perioperative period (*not graded*).
37. We suggest antiparasitic drugs with corticosteroid therapy following shunt insertion to decrease subsequent shunt failure in patients in whom surgical removal of isolated intraventricular cysts is not possible (*weak, low*), but neither after successful removal of intraventricular cysts (*weak, low*). *Remark:* Note that intraventricular cysts may be accompanied by other lesions with indications for antiparasitic therapy.

## RECOMMENDATIONS FOR SUBARACHNOID NEUROCYSTICERCOSIS

### XXII. What is the role of medical therapy in subarachnoid neurocysticercosis (SAN) in the basilar cisterns or Sylvian fissures?

#### Recommendations

38. We recommend that patients with subarachnoid cysts be treated with antiparasitic drugs (*strong, low*).
39. We suggest that antiparasitic therapy be continued until there is radiologic resolution of viable cysticerci on MRI and resolution of other evidence of cysticerci (*weak, low*). Responses often require prolonged therapy, which can last for more than a year.



40. We recommend anti-inflammatory therapy (such as high-dose corticosteroids) for subarachnoid NCC initiated prior to antiparasitic drugs (*strong, moderate*).
41. We suggest that methotrexate be considered as a steroid-sparing agent in patients requiring prolonged courses of anti-inflammatory therapy (*weak, low*).

### XXIII. What is the role of neurosurgery in SAN?

#### Recommendations

42. We recommend that patients with hydrocephalus from subarachnoid NCC be treated with shunt surgery in addition to medical therapy (*strong, low*).
43. We suggest that some patients may benefit from surgical debulking over shunt surgery alone (*weak, low*).

## RECOMMENDATIONS FOR SPINAL NEUROCYSTICERCOSIS

### XXIV. How is spinal neurocysticercosis (SN) best treated?

#### Recommendations

44. We recommend corticosteroid treatment for patients with SN with evidence of spinal cord dysfunction (eg, paraparesis or incontinence) or as adjunctive therapy along with antiparasitic therapy (*strong, moderate*).
45. We suggest that both medical (antiparasitic drugs plus anti-inflammatory drugs) and surgical approaches be considered for SN (*weak, low*). *Practice statement:* There are anecdotes of good responses of spinal neurocysticercosis to medical and/or surgical therapy. However, there are no good data supporting one approach over the other. We suggest that management of spinal NCC should be individualized based on symptoms, location of the cysticerci, degree of arachnoiditis, and surgical experience. Recommendations for antiparasitic drugs, reimaging, and follow-up of SAN should also be considered for subarachnoid SN.

## RECOMMENDATIONS FOR MANAGEMENT OF OCULAR CYSTICERCOSIS

### XXV. What is the optimal management of ocular cysticercosis?

#### Recommendation

46. We suggest that intraocular cysticerci be treated with surgical removal rather than with antiparasitic drugs (*weak, low*).

## RECOMMENDATIONS FOR THE TREATMENT OF SPECIAL POPULATIONS

### XXVI. Should children be managed differently than adults?

#### Recommendation

47. There is no evidence that management of NCC in children should be different than in adults with the same form

of disease (*strong, moderate*). Dosing should be weight based.

### XXVII. Should management be different in pregnant women?

#### Recommendation

48. We suggest that antihelminthic therapy be deferred until after pregnancy (*weak, low*). *Remarks:* Pregnant patients with elevated intracranial pressure need to be aggressively managed as they would be if not pregnant. Corticosteroids can be used in pregnancy when necessary. The use of antiepileptic drugs should take into account altered pharmacokinetics and potential teratogenicity. Phenobarbital and valproic acid are known to have high rates of teratogenicity. Antihelminthic drugs are rarely required emergently and their use can usually be deferred until after delivery. Methotrexate is teratogenic and should be avoided.

#### Notes

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**Potential conflicts of interest.** The following list is a reflection of what has been reported to IDSA. To provide thorough transparency, IDSA requires full disclosure of all relationships, regardless of relevancy to the guideline topic. Evaluation of such relationships as potential conflicts of interest (COI) is determined by a review process that includes assessment by the SPGC Chair, the SPGC liaison to the development panel, the Board of Directors liaison to the SPGC, and, if necessary, the COI Task Force of the Board. This assessment of disclosed relationships for possible COI will be based on the relative weight of the financial relationship (ie, monetary amount) and the relevance of the relationship (ie, the degree to which an association might reasonably be interpreted by an independent observer as related to the topic or recommendation of consideration). The reader of these guidelines should be mindful of this when the list of disclosures is reviewed. For activities outside of the submitted work, A. C. W. has received royalties from UpToDate. For activities outside of the submitted work, A. H. has served as a member of a data and safety monitoring board for Neuropace. For activities outside of the submitted work, H. G. has received research grants from the National Institute of Neurological Disorders and Stroke and the Fogarty International Center of the National Institutes of Health. All other authors report no potential conflicts. All authors have submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. Conflicts that the editors consider relevant to the content of the manuscript have been disclosed.

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