

Table 76: Tests 1, 2 and 4. Index tests (APRI, Forn's score and Transient Elastography) versus clinical and ultrasound cirrhosis definition to detect cirrhosis in a population with CFLD (practice guideline defined) †

Number of studies (Reference)	Study design	N	Risk of bias	Inconsistency	Indirectness	Imprecision	Sensitivity % (95% CI)	Specificity % (95% CI)	Positive likelihood ratio (95% CI)	Negative Likelihood ratio (95% CI)	AUROC	Quality
Test 1. APRI using a cut off of 0.344 in a population of adults with CFLD												
1 (Karlas 2012)	Cohort study	14	no serious risk of bias	no serious inconsistency	no serious indirectness	very serious imprecision ^a	83.3 (95% CI: 45.0-98.5)*	87.5 (95% CI: 58.8-98.9)*	6.67 (95% CI: 1.09-88.5)*	0.19 (95% CI: 0.02-0.94)*	0.88 (95% CI: 0.59-0.99)	LOW
Test 2. Forn's score using a cut off of 4.059 in a population of adults with CFLD												
1 (Karlas 2012)	Cohort study	14	no serious risk of bias	no serious inconsistency	no serious indirectness	very serious imprecision ^a	66.7 (95% CI: 30.1-75.0)*	94.1 (95% CI: 68.3-100)*	11.3 (95% CI: 0.95-6684670)*	0.35 (95% CI: 0.25-1.02)*	0.85 (95% CI: 0.57-0.98)	LOW
Test 4. Transient elastography using a cut off of 4.4kPa in a population of adults with CFLD												
1 (Karlas 2012)	Cohort study	14	no serious risk of bias	no serious inconsistency	no serious indirectness	very serious imprecision ^a	92.3 (95% CI: 56.2-100)*	75 (95% CI: 45.7-81.2)*	3.69 (95% CI: 1.04-5.33)*	0.10 (95% CI: 0-0.96)*	0.88 (95% CI: 0.59-0.99)	LOW

Abbreviations: AUROC: area under the ROC curve; APRI: Aspartate aminotransferase to Platelets-Ratio-Index; CFLD: cystic fibrosis related disease; CI: confidence interval
†Diagnosis of CFLD (Sokol 1999, Colombo 2002) if at least 2 of the following conditions present on at least 2 consecutive examinations spanning a 1-year period: (1) Ultrasound confirmed hepatomegaly;(2) elevated serum liver enzyme levels of ALT, AST, AP, or GGT;(3) ultrasound abnormalities other than hepatomegaly (i.e., increased, heterogeneous echogenicity, nodularity, irregular margins, splenomegaly). Liver cirrhosis: distinct ultrasonographic signs (i.e. coarse nodularity, presence of portal hypertension and rarefaction of peripheral portal veins) and clinical signs (e.g. esophageal varices, splenomegaly)

* Calculated by the NGA technical team from data available in the study report

a. 95% confidence interval for sensitivity was very wide (width ≥30 percentage points)