

Table 69: Tests 8 & 13. Index tests (Ultrasound and Transient elastography) versus Clinical CFLD definition† to detect CFLD

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 8. Ultrasound (cut off value Williams score ≥ 4) in a population of adults and children												
1 (Witters 2009)	Cohort study	66	no serious risk of bias	no serious inconsistency	no serious indirectness	very serious imprecision ^a	66.7 (95% CI: 25.0-93.9)*	66.7 (95% CI: 62.5-69.4)*	2.0 (95% CI: 0.67-3.07)*	0.50 (95% CI: 0.09-1.2)*	0.77 (95% CI: 0.51-1.02)	LOW
Test 13. Transient elastography using Fibroscan (Age-specific cut-off values at 5.63kPa for <12 years and 6.50kPa for ≥ 12 years) in a population of adults and children												
1 (Witters 2009)	Cohort study	66	no serious risk of bias	no serious inconsistency	no serious indirectness	very serious imprecision ^a	83.3 (95% CI: 38.7-99.1)*	85.0 (95% CI: 80.5-86.6)*	5.6 (95% CI: 2.0-7.4)*	0.20 (95% CI: 0.01-0.76)*	0.93 (95% CI: 0.85-1.01)	LOW

Abbreviations: AUROC: area under the curve; CFLD: cystic fibrosis liver disease; CI: confidence interval; kPa: kilopascal

†Diagnosis of CFLD according to the presence or absence of hepatomegaly or splenomegaly determined by clinical examination

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

a. 95% confidence interval for sensitivity was very wide (width $\geq 30\%$)