

Table 70: Tests 9 & 14. Index tests (Ultrasound and Transient elastography) versus Biochemical 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 9. Ultrasound (cut off of Williams score ≥ 4) in a population of adults and children												
1 (Witters 2009)	Cohort study	6 6	no serious risk of bias	no serious inconsistency	no serious indirectness	very serious imprecision ^a	50.0 (95% CI: 14.3-85.6)*	66.7 (95% CI: 63.1-70.2)*	1.5 (95% CI: 0.39-2.88)*	0.75 (95% CI: 0.21-1.36)*	0.62 (95% CI: 0.40-0.84)	LOW
Test 14. Transient elastography using Fibrosan (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	6 6	no serious risk of bias	no serious inconsistency	no serious indirectness	very serious imprecision ^a	50.0 (95% CI: 14.5-85.3)*	83.3 (95% CI: 79.8-86.9)	3.0 (95% CI: 0.72-6.5)*	0.60 (95% CI: 0.17-1.07)*	0.78 (95% CI: 0.61-0.95)	LOW

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

†Diagnosis of CFLD was defined as persistently elevated results (3–6 months, 1.5 times age-dependent upper limit of normal) for 2 of these liver tests: AST, ALT, alkaline phosphatase, bilirubin and gamma-GT.

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

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