**Table 78: Tests 1 to 3. Index tests (APRI, Forn's score, transient elatography) versus clinical definition to detect portal hypertension†**

| 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 at a cut off of ≥ 0.49 in a population of adults** | | | | | | | | | | | | |
| 1(Kitson 2013) | Case control study | 50 | no serious risk of bias | no serious inconsistency | no serious indirectness | no serious imprecision | 87.5 (95% CI: 52.0-99.3)* | 92.9 (95% CI: 86.1-95.1)* | 12.3 (95% CI: 3.74-20.3)* | 0.14 (95% CI: 0.01-0.56)* | 0.97 (95% CI: 0.93-1.00) | LOW |

| 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. Subgroup analysis: APRI at a cut off of ≥ 0.49 in a population of adults with CFLD*** | | | | | | | | | | | | |
| 1(Kitson 2013) | Case control study | 25 | no serious risk of bias of bias | no serious inconsistency | no serious indirectness | no serious imprecision | 87.5 (95% CI: 54.8-98.9)* | 94.1 (95% CI: 78.7-99.5)* | 14.9 (95% CI: 2.6-189.4)* | 0.13 (95% CI: 0.01-0.58)* | 0.98 (95% CI: 0.93-1.00) | LOW |
| **Test 2. Forn's at a cut off of ≥ 0.68 in a population of adults** | | | | | | | | | | | | |
| 1(Kitson 2013) | Case control study | 50 | no serious risk of bias | no serious inconsistency | no serious indirectness | no serious imprecision | 87.5 (95% CI: 50.7-99.3)* | 85.7 (95% CI: 78.7-88.0)* | 6.13 (95% CI: 2.38-8.26)* | 0.15 (95% CI: 0.01-0.63)* | 0.93 (95% CI: 0.85-1.00) | LOW |
| ***Test 2. Subgroup analysis: Forn's score at a cut off of ≥ 0.68 in a population of adults with CFLD*** | | | | | | | | | | | | |
| 1(Kitson 2013) | Case control study | 25 | no serious risk of bias | no serious inconsistency | no serious indirectness | no serious imprecision | 87.5 (95% CI: 53.2-99.3)* | 82.4 (95% CI: 66.2-87.9)* | 5.0 (95% CI: 1.6-8.2)* | 0.15 (95% CI: 0.01-0.71)* | 0.93 (95% CI: 0.82-1.00) | LOW |
| **Test 3. Transient elastography at a cut off of ≥ 8.9 kPa in a population of adults** | | | | | | | | | | | | |
| 1(Kitson 2013) | Case control study | 50 | no serious risk of bias | no serious inconsistency | no serious indirectness | no serious imprecision | 87.5 (95% CI: 51.4-99.3)* | 90.5 (95% CI: 83.6-92.7)* | 9.19 (95% CI: 3.14-13.66)* | 0.14 (95% CI: 0.01-0.58)* | 0.96 (95% CI: 0.92-1.00) | LOW |
| ***Test 3. Subgroup analysis: Transient elastography at a cut off of ≥ 8.9 kPa in a population of adults with 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 |
|---|---|---|---|---|---|---|---|---|---|---|---|---|
| 1(Kitson 2013) | Case control study | 25 | no serious risk of bias | no serious inconsistency | no serious indirectness | no serious imprecision | 87.5 (95% CI: 52.9-99.3)* | 76.5 (95% CI: 60.2-82.0)* | 3.7 (95% CI: 1.33-5.53)* | 0.16 (95% CI: 0.01-0.78)* | 0.91 (95% CI: 0.79-1.00) | LOW |

*Abbreviations: APRI Aspartate aminotransferase to Platelets-Ratio-Index; AUROC: area under the ROC curve; CFLD: cystic fibrosis liver disease; CI: confidence interval; kPa: kilopascal*

*†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 rarefication of peripheral portal veins) and clinical signs (e.g. esophageal varices, splenomegaly). Portal hypertension: platelet count <140x109/L, splenomegaly, presence of porto-systemic collateral veins, portal diameter >13mm, or ascites*

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