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| **About the Measure**  |
| **Domain:** | Sickle Cell Disease Curative Therapies |
| **Measure:** | Determination of Liver Fibrosis by Transient Elastography |
| **Definition:** | Fibrosis staging defines the degree of liver scarring and indicates how far disease has progressed. Liver fibrosis is a wound healing response typically resulting from chronic injury, in which the connective tissue matrix is overproduced, inefficiently degraded, or both. Advanced liver fibrosis may lead to the adverse clinical consequences of portal hypertension.There is a high frequency of liver disease in sickle cell disease (SCD) patients who are candidates for curative therapies. Mechanisms of liver injury in SCD include iron overload, vaso-occlusive crises, biliary injury, immune injury, and viral hepatitis (Berry et al., 2007; Feld et al., 2015; Jitraruch et al., 2017; Theocharidou et al., 2019). Advanced liver fibrosis may increase the risk or preclude the use of some curative therapies. |

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| **About the Protocol** |
| **Description of Protocol:** | This protocol provides an overview of performing vibration controlled transient elastography of the liver.  |
| **Protocol:** | **Description of Determining Liver Fibrosis by Transient Elastography (FibroScan®)**The FibroScan® technology produces a mechanically induced elastic shear wave that is propagated in the liver. An ultrasound probe with a vibration transducer measures the speed of the wave. The speed of the shear wave is used to estimate liver stiffness, which is correlated with fibrosis. A full description of the process for determining Liver Fibrosis by Transient Elastography can be found in the Materials and Methods section of Sandrin et al., 2003. This includes the details of the FibroScan® apparatus, propagation of the low-frequency excitation, estimation of liver elasticity, probe placement, measurement acquisition and statistical procedures.  |
| **Participant:** | Adults, ages 18 and older |
| **Source:** | Sandrin, L., Fourquet, B., Hasquenoph, J. M., Yon, S., Fournier, C., Mal, F., Christidis, C., Ziol, M., Poulet, B., Kazemi, F., Beaugrand, M., & Palau, R. (2003). Transient elastography: A new noninvasive method for assessment of hepatic fibrosis. *Ultrasound in Medicine & Biology, 29*(12), 1705–1713.  |
| **Language of Source:** | English  |
| **Personnel and Training Required:** | Vibration controlled transient elastography by Fibroscan® should be administered by appropriately trained personnel and interpreted by clinicians (e.g., radiologists, hepatologists, hematologists) with relevant expertise. |
| **Equipment Needs:** | FibroScan® medical device  |
| **Protocol Type:** | Complex instrumentation-based assessment |
| **General References:** | Berry, P. A., Cross, T. J., Thein, S. L., Portmann, B. C., Wendon, J. A., Karani, J. B., Heneghan, M. A., & Bomford, A. (2007). Hepatic dysfunction in sickle cell disease: A new system of classification based on global assessment. *Clinical Gastroenterology and Hepatology, 5*(12), 1469–1476. Costa, P., Rudolph, B., Kogan-Liberman, D., Manwani, D., Silver, E. J., & Ovchinsky, N. (2020). Liver stiffness measurement by vibration controlled transient elastography does not correlate to hepatic iron overload in children with sickle cell disease. *Journal of Pediatric Hematology/Oncology*, *42*(3), 214–217. Feld, J. J., Kato, G. J., Koh, C., Shields, T., Hildesheim, M., Kleiner, D. E., Taylor, J. G., 6th, Sandler, N. G., Douek, D., Haynes-Williams, V., Nichols, J. S., Hoofnagle, J. H., Jake Liang, T., Gladwin, M. T., & Heller, T. (2015). Liver injury is associated with mortality in sickle cell disease. *Alimentary Pharmacology and Therapeutics, 42*(7), 912–921. Foucher, J., Chanteloup, E., Vergniol, J., Castéra, L., Le Bail, B., Adhoute, X., Bertet, J., Couzigou, P., & de Lédinghen, V. (2006). Diagnosis of cirrhosis by transient elastography (FibroScan): A prospective study. *Gut*, *55*(3), 403–408. Jitraruch, S., Fitzpatrick, E., Deheragoda, M., Deganello, A., Mieli-Vergani, G., Height, S., Rees, D., Hadzic, N., & Samyn, M. (2017) Autoimmune liver disease in children with sickle cell disease. *Journal of Pediatrics, 189*, 79–85.Pinto, V. M., Gianesin, B., Balocco, M., Bacigalupo, L., & Forni, G. L. (2017). Noninvasive monitoring of liver fibrosis in sickle cell disease: Longitudinal observation of a cohort of adult patients. *American Journal of Hematology*, *92*(12), E666–E668. Theocharidou, E., & Suddle, A.R., (2019). The liver in sickle cell disease. *Clinical Liver Disease, 23*(2), 177–189.  |