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| **About the Measure**  |
| **Domain** | Sickle Cell Disease Curative Therapies |
| **Measure** | Determination of Liver Fibrosis by Magnetic Resonance Elastography (MRE) |
| **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 exists 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 specifications for determining liver fibrosis by magnetic resonance (MR) elastography. Quantitative measures are generated by the imaging software and used to determine the presence of liver fibrosis. |
| **Protocol** | **Description of Quantification of Liver Fibrosis by Magnetic Resonance Elastography (MRE)**A description of MRE studies for the determination of liver fibrosis can be found in Yin et al. 2007. Briefly, MR elastography is performed on a 1.5-T whole-body scanner, using a whole body coil. Low-amplitude mechanical waves at 60 Hz are generated in the abdomen using an acoustic driver on the anterior body wall. The total data acquisition time is 40 seconds. Images are quantified to display shear stiffness (MR elastograms) by propagating shear waves with a local frequency estimation inversion algorithm.The supplementary material from Yin et al. 2007 provides details of the MR elastography protocol, processing of elastograms, measurement of steatosis using MR, and optimization of the driver used in MR elastography. |
| **Participant** | Age 18 years or older |
| **Source** | Yin, M., Talwalkar, J. A., Glaser, K. J., Manduca, A., Grimm, R. C., Rossman, P. J., Fidler, J. L., & Ehman, R. L. (2007). Assessment of hepatic fibrosis with magnetic resonance elastography. *Clinical Gastroenterology and Hepatology*, *5*(10), 1207–1213. |
| **Language of Source** | English |
| **Personnel and Training Required** | A trained magnetic resonance imaging (MRI) technician is required to administer the MRI, and MRIs must be interpreted (“read”) by a trained radiologist, cardiologist, or other medical doctor. |
| **Equipment Needs** | A magnetic resonance imaging (MRI) machine at 1.5 Tesla with multiple-echo, gradient echo sequence for R2\*/T2\* evaluation. |
| **Protocol Type** | Imaging 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. 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. 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.Mariappan, Y. K., Glaser, K. J., & Ehman, R. L. (2010). Magnetic resonance elastography: A review. *Clinical Anatomy*, *23*(5), 497–511.Muthupillai, R., Lomas, D. J., Rossman, P. J., Greenleaf, J. F., Manduca, A., & Ehman, R. L. (1995). Magnetic resonance elastography by direct visualization of propagating acoustic strain waves. *Science*, *269*(5232), 1854–1857.Theocharidou, E., & Suddle, A.R., (2019). The liver in sickle cell disease. *Clinical Liver Disease, 23*(2), 177–189. Trout, A. T., Sheridan, R. M., Serai, S. D., Xanthakos, S. A., Su, W., Zhang, B., & Wallihan, D. B. (2018). Diagnostic performance of MR elastography for liver fibrosis in children and young adults with a spectrum of liver diseases. *Radiology*, *287*(3), 824–832.Venkatesh, S. K., Yin, M., & Ehman, R. L. (2013). Magnetic resonance elastography of liver: Technique, analysis, and clinical applications. *Journal of Magnetic Resonance Imaging*, *37*(3), 544–555.Wang, Q. B., Zhu, H., Liu, H. L., & Zhang, B. (2012). Performance of magnetic resonance elastography and diffusion-weighted imaging for the staging of hepatic  |