**SUPPLEMENTAL FIGURE LEGENDS**

**Figure S1** *Mdr2* deficiency results in cholestatic liver injury and tumor development. *Mdr2-/-*and WT mice at the age of 8 and 52 weeks were included in this analysis. (A) Serum aspartate aminotransferase levels (AST).(B) The liver weight *versus* body weight ratio. (C) Serum cholesterol levels.(D) Representative sirius red staining of liver paraffin sections are shown. (E) Quantification represents positive staining as percentage of area fraction using ImageJ© software. (F) Macroscopic appearance of 52 week-old *Mdr2-/-*livers showing large tumors on liver surface. (G) *Mdr2-/-* mice display well-differentiated hepatocellular tumors with focal atypia and small group necrosis. (H) Histological scoring of *Mdr2-/-* livers. (I) Data table of histological scoring. Data are expressed as the mean ± SD from 12 to 15 mice per group and considered significant if \*\* *p*<0.01, \*\*\* *p*<0.001.

**Figure S2** An increased inflammatory response is found in *Mdr2-/-* livers. (A) Immunohistochemical staining against KI67 in livers of 52 week-old *Mdr2-/-* and WT mice. (B) CD45 immunohistochemical staining and quantification in livers of *Mdr2-/-* and WT mice at 8 and 52 weeks age. (C) Liver mRNA expression of MCP1 as determined by qRT-PCR from *Mdr2-/-* and WT mice at 8 and 52 weeks of age. (D) FACS analysis of monocyte-derived macrophages (defined as F4/80low CD11bhi) in 8 week-old WT and *Mdr2-/-* livers. (E) F4/80 immunohistochemical staining in livers of *Mdr2-/-* and WT mice at 8 and 52 weeks of age. (F) mRNA expression of CD68 and F4/80 as determined by qRT-PCR in 8 week-old WT and *Mdr2-/-* livers. Data are expressed as the mean ± SD from 6 to 8 mice per group and considered significant if \* *p*<0.05, \*\* *p*<0.01, \*\*\* *p*<0.001.

**Figure S3** Apoptotic cell death is activated in *Mdr2-/-* livers, but caspase-8 in liver parenchymal cells has no significant impact on disease progression.*Mdr2-/-*and WT livers of 8 and 52 week-old mice were included in this analysis. (A) Caspase-3 and caspase-8 enzyme activities were analyzed. (B) Expression of cleaved caspase-9 protein was determined by western blot analysis. GAPDH was used as loading control. (C) Quantification of RIP1 and RIP3 western blot bands. (D) Bcl2 and Bax mRNA expression were determined by qRT-PCR. Data are expressed as the mean ± SD from 6 to 8 mice per group and considered significant if \*\* *p*<0.01, \*\*\* *p*<0.001.

**Figure S4** Caspase-8 in liver parenchymal cells has no significant impact on disease progression. (A) Liver weight *versus* body weight ratios are shown of 26 week-old *Mdr2-/-*/Casp8hepa and *Mdr2-/-*/Casp8f/f mice. (B) Sirius red staining of 26 week-old *Mdr2-/-*/Casp8hepa and *Mdr2-/-*/Casp8f/f livers was performed and representative images of liver paraffin sections are shown (left panel). Quantification represents positive staining as percentage of area fraction using ImageJ© software (right panel). Each dot represents the value of one independent view field. (C) NLRP3, IL-1β score and MELD score plotted against the grade of fibrosis observed in PSC patients (n=8) (D) Quantification of pro-IL-1β, IL-1β and NLRP3 western blot bands performed in liver tissue. (E) Microbiota composition (family level) of *Mdr2-/-* and WT mice. Every bar represents the community structure (caecal content) of one mouse. (F) Pearson correlation analysis between Lachnospiraceae OTU\_69 abundance and serum ALT levels in *Mdr2-/-* mice. (G) NMDS-graphs based on Bray-Curtis dissimilarity shows clear separation of *Mdr2-/-* and WT microbiota; Litters and Cages are shown in different colors. (H) PCoA-graphs based on UniFrac distances of *Mdr2-/-* and WT microbiota; Litters and Cages are shown in different colors. Data are expressed as the mean ± SD from 5 to 12 mice per group and considered significant if \*\* *p*<0.01, \*\*\* *p*<0.001.

**Figure S5** *Mdr2-/-* intestines display intestinal barrier impairment.Colon and ileum samples of *Mdr2-/-* and WT mice at 8 and 52 weeks of age were included in this analysis.(A) Representative H&E stainings of paraffin colon and (B) ileum tissue sections. (C) Quantification of ZO-1 western blot bands were performed on colon (left panel) and ileum (right panel) tissue. (D) Thickness of the mucus layer measured by histological analyses after mucin-2 immunofluorescence staining using AxioVision® software. Data are expressed as the mean ± SD from 6 to 8 mice per group and considered significant if \* *p*<0.05, \*\* *p*<0.01.

**Figure S6** *Mdr2-/-* intestines display intestinal inflammation.Colon and ileum samples of *Mdr2-/-* and WT mice at 8 and 52 weeks of age were included in this analysis.(A) Representative CD45, (B) CD11b and (C) F4/80 immunohistochemical stainings on paraffin colon and ileum sections are shown. (D) F4/80 and (E) TNFα mRNA expression were determined by qPT-PCR in Colon and Ileum of WT and *Mdr2-/-* mice. (F) Quantification of NLRP3, pro-IL-1β and IL-1β western blot bands performed in colon tissue. Data are expressed as the mean ± SD from 3 to 8 mice per group and considered significant if \* *p*<0.05, \*\* *p*<0.01.

**Figure S7** FMT of *Mdr2-/-*microbiota induces intestinal dysbiosis in recipient WTFMT(*Mdr2-/-*) mice. 8 week-old *Mdr2-/-* and WT mice +/- IDN 7314 treatment were included in this analysis. (A) Serum GLDH and AP levels. (B) Immunofluorescence staining against ZO-1 demonstrates reduced expression ZO-1 in WT mice receiving *Mdr2-/-* microbiota via oral gavage (WTFMT(*Mdr2-/-*)). (C) CD45 immunohistochemical staining on paraffin liver sections and staining quantification. (D) Intestinal microbiota composition (family level) of WTFMT(WT) and WTFMT(*Mdr2-/-*) mice before (d0) and after (d7) repetitive FMT as well as WT and *Mdr2-/-* donor mice. (E) Microbiota community structure of WT donor mice vs. Mdr2-/- donors. (F) Microbiota community structure of WTFMT(*Mdr2-/-*) vs. WTFMT(WT) mice after one week of repetitive FMT. (G) Microbiota of WTFMT(*Mdr2-/-*) vs. *Mdr2-/-* donors after FMT. Data are expressed as the mean ± SD from 3 to 5 mice per group.

**Figure S8** IDN-7314 treatment reshapes intestinal microbiota composition and inhibits liver inflammasome activation in *Mdr2-/-* mice. 8 week-old *Mdr2-/-* and WT mice +/- IDN 7314 treatment were included in this analysis. (A) Quantification of cleaved caspase-8 (P43 and P18) and (B) cleaved caspase-3 western blot bands performed on liver tissue. (C)Bar chart based on permutational multivariate analysis of variance (ADONIS) presenting percentage of variability of the gut microbiota explained by the factor “IDN 7314 treatment” in caecal content.(D) Quantification of NLRP3, IL-1β and pro-IL-1β of western blot bands. (E) CD11b immunofluorescence staining (left panel) and quantification (right panel) in livers of *Mdr2-/-* and WT mice. Data are expressed as the mean ± SD from 6 to 6 mice per group and considered significant if \*\* *p*<0.01, \*\*\* *p*<0.001.

**METHODS**

**Fecal microbiota transfer**

For microbiota modulation experiments (fecal microbiota transfer, FMT), WT mice were treated for 1 week three times/week (Monday-Wednesday and Friday) via oral gavage with 200ul of fecal dilution. To prepare this dilution, per mouse 20mg of freshly harvested stool (immediately upon defecation) was collected from a group of 4 *Mdr2-/-* donor and 3 WT donor mice (all mice between 10-13 weeks old). Stool pellets were pooled and then vortexed for 3 min in 100µl/20mg(stool) anaerobic PBS to dilute it almost entirely. Next, samples were gently centrifuged for 2 minutes at 50g to allow stool particulate to settle. Supernatant was collected and diluted 1:1 with anaerobic PBS. 200µl of this suspension was transplanted by oral gavage into recipient mice (8 weeks old, WT littermates).

**qRT–PCR**

Total RNA was extracted with Trizol Reagent (Life Technologies) and reverse transcription was performed using an Omniscript kit (Qiagen) according to the manufacturer’s protocol. For the assembling of real time PCR reactions SYBR® GreenER qPCR SuperMix (Invitrogen) was used according to manufacturer’s recommendations. Primer sequences are available upon request. The Real-Time PCR System (AB 7300) was used. Expression of mRNA was calculated using the 2−ΔΔCT Method, which determines the relative quantification of a target gene in comparison to the GAPDH gene.

**Western blot**

The protein samples (1µg/µl) were separated electrophoretically on pre-cast 4-12% polyacrylamide gel (Invitrogen) in MOPS running buffer at 120 V for approximately 1.5 hours. For immunological detection, the separated proteins in the gel were transferred to a nitrocellulose membrane. Successful protein transfer was confirmed by Ponceau Red staining. For immunological detection, the non-specific binding sites were blocked for 1 hour in 5% non-fat dry milk diluted in TBS-Tween (TBST). The membrane was incubated overnight at 4°C in TBST with primary antibody at optimized dilution. After being washed the membrane was incubated with the horseradish peroxidase (HRP)-conjugated secondary antibody diluted 1:5000 in 5% non-fat dry milk diluted in TBST for 1 hour at room temperature. After incubation, the membrane was washed in TBST to wash away the non-bound antibody and then incubated in ECL Substrate (Pierce) for 5 min. The membrane was exposed to a LAS mini 4000 developing machine (Fuji) until specific signals were detectable. In this study, the following antibodies were used: IL-1β (Abcam, AB9722), Caspase-1(Canta Cruz, SC-514),Caspase-8 (Enzo life sciences, ALX-804-447), Cleaved caspase-8 (Cell Signaling, D5B2), Cleaved caspase-3 (Cell Signaling, #9661), Cleaved caspase-9 (Cell Signaling, #9509S), Collagen Iα1 (BioTrend, BT-5014-10), RIP-1 (BD, 610459), RIP3 (Prosci, 2283), NLRP3 (Abcam, Ab4207; Adipo Gen, AG-20B-0014), P-MLKL (Abcam, AB196436), P-P65 (Cell Signaling, 3033S), ZO-1 (Abcam, Ab96587), GAPDH (Bio-RAD, MCA4739), β-actin (Sigma, A2066).

**Caspase activity assay**

Caspase-3 and caspase-8 are key enzymes for induction of the apoptotic cascade which participate in the cleavage of protein substrates. According to the manufacturer’s protocol, per reaction tube 487.5 µl mastermix and 12.5 µl protein were mixed, followed by pipetting 200 µl of the mixture in duplicate into a 96-well-cellstar plate and incubating at 37°C. Mastermix with lysis buffer was taken as blank. AFC-release was measured in 30 min intervals and stopped after 3 hours. Caspase-3 substrate was Ac-DEVD-AFC (Enzo life sciences, ALX-260-032-M005), and caspase-8 substrate was Ac-LETD-AFC (Enzo life sciences, ALX-260-118-M005). Measurements were performed with a spectrofluorometer at 400 nm excitation and 505 nm emission wavelengths.

**Immunofluorescence staining**

ZO-1 immunofluorescence staining was performed on cryopreserved 5 µm intestine sections of mice. The sections were dried in air for 15-20 minutes, fixed with 4% PFA for 8-10 min in room temperature (RT) and washed with PBS for 3 x 5 min. PBS containing 5% goat serum was used for blocking for 30 min followed by an incubation with ZO-1 antibody (Abcam, Ab96587) in blocking solution at optimized dilutions at 4°C overnight in a humidified chamber. Next day slides were washed with PBS for 3 x 5 min and incubated with Alexa Fluor 488 anti-rabbit (Invitrogen, A-11008) in blocking buffer for one hour at RT in humidified chamber. Slides were washed with PBS for 3 x 5 min. Nuclei were counterstained with DAPI mounting medium (Dako).

The terminal deoxynucleotidyl transferase-mediated deoxyuridine triphosphate nick-end labeling (TUNEL) assay (Roche) was performed on liver cryosections. First liver cryosections were fixed with 4% PFA for 45 min and rehydrated with PBS for 3 x 10 min. After treating with 3% H2O2 in methanol for 10 min, the sections were permeabilized with 0.1% Triton and 150 mM Na-citrate for 2 min at 4°C. Slides were washed with PBS and stained with TUNEL mix (1 μl enzyme, 9 μl TUNEL dilution buffer, 90 μl buffer) in a humidified chamber overnight at 4°C. After 3 x 10 min washing with PBS stained sections were mounted with DAPI mounting medium (Dako). The percentage of TUNEL positive cells was counted and set in percentage relationship to the total cells from the same field.

Mucus staining was performed according to an established protocol(1). Colon tissue sections containing fecal pellets were fixed using the Carnoy fixation method (60% absolute methanol, 30% chloroform, 10% glacial acetic acid) for 48h followed by embedding in paraffin. Paraffin embedded sections were rehydrated with xylene and decreasing percentages of ethanol. Sections were blocked with 10% goat serum 20 min and further incubated overnight with anti-Muc2 primary antibody (1:500 diluted, SC-15334, Canta Cruz) at 4°C in a humid atmosphere. Slides were washed twice with PBS 5 min x 3 times, and the secondary antibody (1:200 diluted) was applied for 1 hour at RT in humidifying box. Nuclei were counterstained with DAPI in mounting medium.

**Immunohistochemistry staining**

Immunohistochemical stainings against CD45 (BD, 550539), CD11b (BD, 550282), F4/80 (AbD serotec, MCA497GA), cleaved Caspase-3 (Cell Signaling, #9661), cleaved Caspase-8 (Cell Signaling, #9429S) and NLRP3 (Adipo Gen, AG-20B-0014) were performed on paraffin-embedded sections, as describe previously(2). The sections were blocked with peroxidase-conjugated avidin-biotin method (ABC Reagent Kit, Vector). To block unspecific binding sites slides were further incubated for 30 min at RT in 50% FCS + 50% PBS containing 1% BSA. Slides were then incubated with primary antibody in blocking solution at optimized dilutions at 4°C overnight. Next day slides were washed twice with PBS and the secondary antibodies (1:200 diluted in PBT) were applied for 45 min at RT. To make the visualization of the signal, the enzyme substrate 3, 3'-diaminobenzidine (DAB) Substrate-Kit (Vector) was used.

**Sirius red**

To study the development of liver fibrosis, paraffin embedded tissue sections were stained with Sirius red, as described previously(3). The slides were dewaxed, rehydrated and incubated in Sirius red staining solution for 45 min. The staining was fixed in 0.5% Acetic Acid. Afterwards the sections were dehydrated and mounted using Roti®-Histokitt. The positive area fraction was quantified via ImageJ©.

**Flow cytometry analysis of intrahepatic leukocytes**

Immune cells isolated from livers of mice were incubated with blocking buffer 20min to block the unspecific binding sites of cell surface and then stained with fluorochrome-conjugated antibodies against Ly6G, CD11b, CD11c, F4/80, Gr1.1 and CD45 (1: 200), all from eBioscience (Frankfurt, Germany). All samples were acquired by flow cytometry (FACS Canto II; BD Biosciences, Heidelberg, Germany) and analyzed using the Flowjo software (Ashland, OR, USA).

**Bile acid analysis**

Bile acids in caval venous blood were analyzed using ultra-performance liquid chromatography-tandem mass spectrometry (UPLC-MS/MS, Wallenberglab laboratory at Sahlgrenska University Hospital, Sweden) and quantified employing a combination of deuterium-labeled internal standards and unlabeled standards.

**Intestinal permeability in *vivo***

Isothiocyanate conjugated dextran (FITC-dextran, molecular mass 4.0 kDa, Uppsala, Sweden) dissolved in PBS at a concentration of 200 mg/ml was administered to mice (10 ml/kg body weight) by oral gavage. 4 h after gavage the mice were sacrificed under general anesthesia by isoflurane. Blood samples were collected from inferior vena cava and immediately stored at 4 °C in in the dark. Once blood has been collected in the SST tubes from all the mice, the SST tubes are processed to separate the serum following the manufacturer’s instruction. Add 100 μl of serum to a 96-well microplate in duplicate. Concentration of FITC in serum was determined by spectrophotofluorometry at an excitation wavelength of 485 nm (20 nm band width) and an emission wavelength of 528 nm (20 nm band width), using standard serially diluted FITC-dextran (0, 125, 250, 500, 1,000, 2,000, 4,000, 6,000, 8,000, 10,000 ng/ml). Serum from mice not administered with FITC-dextran was used to determine the background.

**Endotoxin measurement - Limulus amebocyte lysate (LAL) assay**

The Limulus Amebocyte Lysate test (LAL test – Charles River LAL Kit) was used to determine the endotoxin levels in portal serum samples. To avoid external endotoxin contamination only autoclaved endotoxin free tubes and tips were used and the assay was performed in Endosafe 96-well plates. Endotoxin levels were measured in EU/mL where "EU" stands for Endotoxin Units. LPS standard was prepared by diluting 10ng (i.e. 100U) in 10mL of LAL reagent water. This LPS stock was further diluted 1:5 for the highest 20U/mL standard used in the assay, which was then used to prepare the following 12 standards in 1:3 dilutions with LAL reagent water. Serum samples were prepared in endotoxin free tubes and a 1:10 dilution of each sample was made on a microcentrifuge tube rack using 60 µl of sample and adding 540 µl of LAL reagent water, each sample was distributed to 3 endotoxin free tubes containing each 200 µl of the sample and heated for 10 min, 20 min and 30 min, respectively, at 75 °C. Before transfer to the 96-well endosafe plate, all samples were split and half of the samples were spiked with 50µl of 20U/ml LPS-standard. Next samples were pipetted to the 96-well plate, 50 µl of LAL-reagent was quickly added and mixed by tapping the side of the plate several times, then reading promptly was started at 405 nm for 2.5 hours. One EU is equivalent to 100 pg of E. Coli lipopolysaccharide. Results were expressed in EU/ml. In our LPS-spiked samples we measured LPS recovery rates of 85%-100%. Importantly, serum inhibition did not differ between WT and Mdr2-/- mice.

**Microbiota analysis**

16S rRNA analysis. 16S rRNA analysis was performed as previously described in (Roy et al. 2017). Briefly, fresh stool samples of mice were collected and immediately stored at −20°C. DNA was extracted using mechanical disruption (bead-beating) and phenol/chloroform-based purification. In detail, samples were suspended in a solution containing 500µl of extraction buffer (200 mM Tris, 20 mM EDTA, and 200 mM NaCl [pH 8.0]), 200µl of 20% SDS, 500µl of phenol:chloroform:isoamyl alcohol (24:24:1), and 100µl of 0.1 mM zirconia/silica. Samples were homogenized twice with a bead beater (BioSpec) for 2 min. After precipitation of DNA, crude DNA extracts were resuspended in Tris, EDTA (TE) buffer with 100µg/mL RNAse and column-purified to remove PCR inhibitors (BioBasic). Amplification of the V4 region (F515/R806) of the 16S rRNA gene was performed according to previously described protocols ([Caporaso et al., 2011](https://www.cell.com/cell-reports/fulltext/S2211-1247%2817%2931417-1)). Samples were sequenced on an Illumina MiSeq platform (PE250). Sequencing analysis was performed according to previously described computational workflow (4). In brief, obtained reads were assembled, quality controlled and clustered using Usearch8.1 software package (http://www.drive5.com/usearch/). Furthermore, reads were merged using -fastq\_mergepairs –with fastq\_maxdiffs 30 and quality filtering was done with fastq\_filter (-fastq\_maxee 1), minimum read length 200 bp. The OTU clusters and representative sequences were determined using the UPARSE algorithm (5), followed by taxonomy assignment using the Silva database v128 (6) and the RDP Classifier (7) with a bootstrap confidence cutoff of 80%. The OTU absolute abundance table and mapping file were used for statistical analyses and data visualization in the R statistical programming environment (<http://www.rproject.org>) package phyloseq(8). To determine differences in microbial communities between microbiota settings, the LEfSe method was used(9). Ellipses in Figure 8B have been computed in R using the multivariate t-distribution at 0.95 confidence level. (Package ggplot2::stat\_ellipse, The method for calculating the ellipses has been modified from car::ellipse ((Fox and Weisberg, 2011), <https://rdrr.io/cran/ggplot2/man/stat_ellipse.html>). To identify relevant differences in microbiota composition between mice receiving different treatments during FMT experiments, the DESeq2 package was used(10). This package enabled us to compare the OTU count matrices between the different groups and identify bacteria that were differentially regulated between the different groups. The y-axis of the graphs depicts the Log2 fold change of individual OTUs, while the x-axis shows the Log2 mean abundance of the OTU among all samples.

***qRT–PCR***

Liver or intestine tissue was homogenized in 1ml Trizol Reagent (Life Technologies, Carlsbad, CA, USA), then 200 μl chloroform were added to separate the phases, 10 min incubation at RT and 5 min centrifugation at 12000 x g. The aqueous phase was removed and transferred into a new 1.5 ml collecting tube. Afterwards 500 μl isopropanol were added to the tube. After 15 min incubation at RT the RNA was pelleted by 10 min centrifugation at 12000 x g and 4°C. The RNA pellet was washed twice using 70 % ethanol. To transcribe the isolated RNA into cDNA the Omniscipt® RT Kit (200) was used. For reverse transcription 2 μg total RNA were used, and reverse transcription was performed using an Omniscript kit (Qiagen, Venlo, Netherlands) according to the manufacturer’s protocol. Fast SYBR® GreenER Master Mix (Thermo Fisher, Waltham, MA, USA) was used for the assembling of real time PCR reactions according to manufacturer’s recommendations. All the primer sequences are available upon request. For analysis Quant Studio Flex software (Thermo Fisher, Waltham, MA, USA) was used. Expression of mRNA was calculated using the 2−ΔΔCT Method, which determines the relative quantification of a target gene in comparison to the GAPDH gene.

**DNA isolation and measurement of bacterial DNA in liver tissue**

30mg liver tissue was taken from each mouse, homogenized for 10 min and total genomic DNA was extracted directly from the homogenized samples using the peqGOLD Tissue DNA mini Kit according to the manufacturer’s protocol. To check the quality and quantity the NanoDrop spectophotometer was employed. Fast SYBR® GreenER Master Mix (Thermo Fisher, Waltham, MA, USA) was used for the assembling of real time PCR reactions according to manufacturer’s recommendations. All the primer sequences are available upon request. For analysis Quant Studio Flex software (Thermo Fisher, Waltham, MA, USA) was used. Total bacteria, Enterobacteriaceae and Enterococcus were assessed by qPCR of 16s rRNA. qPCR value of bacteria was normalized to the total amount of DNA (in µg) per mg of liver. Additional qPCR of GAPDH was performed for normalization. All gene expression data were expressed as relative to control WT mice.

**Statistical analysis**

For microbiota analyses, statistical analyses were performed using R version 3.4.3 (2017-11-30), (http://www.rproject.org) and the packages ‘phyloseq’ (8), and ‘ggplot2’ (11). The permutational multivariate analysis of variance test (ADONIS) and Analysis of Similarities (ANOSIM) were computed with 999 permutations. For ADONIS tests, a R2 > 0.1 (effect size 10%) and P-value < 0.05 was considered as significant. For ANOSIM tests, a R > 0.2 and P-value < 0.05 was considered as significant. To determine differentially abundant (DA) bacterial families, we used linear discriminant analysis (LDA) effect size (LEfSe)(9) method with Kruskal-Wallis test <0.05 and LDA.

Statistical analyses were performed with GraphPad prism software (version 7.0). For comparisons of 2 groups significance was tested by unpaired two-tailed Student’s t test. In case of more than 2 groups we employed one-way ANOVA followed by Tukey-test with adjusted p-value for multiple comparisons. In case of small sample sizes, two groups were compared using Wilcoxon-Mann-Whitney-Test and in case of more than 2 groups Kruskal-Wallis test with Dunn-Bonferroni-Test. Data were considered significant between experimental groups as: \* p < 0.05, \*\* p < 0.01 or \*\*\* p < 0.001.

**Supplemental Table 1**

**Table 1:** Patient cohort used in the study (n=11). Age and disease activity parameters are given as mean values. “NLRP3” and “IL-1β” indicate the average score observed in patients with the respective grade of hepatic fibrosis (information retrievable for 8 patients). Disease activity parameters were collectable for 10 patients. MELD score: “Model for end-stage liver disease” score. GOT: glutamic-oxaloacetic transaminase. GPT: glutamic-pyruvic transaminase.

|  |  |  |  |
| --- | --- | --- | --- |
|  | **Total** |  |  |
|  |  | *m* | *f* |
| **n** | 11 |  |  |
| Sex |  | 5 | 6 |
| Age | 44,2 | 32 | 54,5 |
|  |  |  |  |
|  |  | *NLRP3* | *IL-1β* |
| **Grade of Liver Fibrosis (n = 8)** |  |  |  |
| I | 2 | 1,5 | 0 |
| II | 2 | 1,5 | 0,5 |
| III | 2 | 2 | 1 |
| IV | 2 | 2,5 | 1 |
|  |  |  |  |
|  |  | *Min* | *Max* |
| **Disease activity (n=10)** |  |  |  |
| MELD Score | 13,5 | 6 | 23 |
| Bilirubin [mg/dl] | 7,22 | 0,25 | 21,8 |
| γ-GT [U/l] | 208 | 25 | 657 |
| GOT [U/l] | 69 | 4 | 162 |
| GPT [U/l] | 72 | 17,4 | 188 |
|  |  |  |  |

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