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Impacto do Desenvolvimento Fetal no Funcionamento Neurocognitivo em Adolescentes com Cardiopatias Congénitas

Sofia Lourenço Sarmento

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Instituto Superior de Ciências da Saúde – Norte

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Sofia Lourenço Sarmento

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Resumo

Objectivo: Avaliar o desempenho neurocognitivo em adolescentes com Cardiopatias Congénitas Cianóticas e Acianóticas e compreender se variáveis neonatais (Índice de Apgar, o Perímetro Cefálico, o comprimento e peso), estão relacionadas com o desempenho neurocognitivo dos adolescentes supra mencionados.

Procedimento: Participaram 48 adolescentes, 26 do género masculino e 22 do género feminino, com idades compreendidas entre os 13 e os 18 anos de idade. A amostra foi dividida em dois grupos, o GC (n=16; 11 do género masculino e 5 do sexo feminino; média das idades=15,69) e o GCC (n=32, 15 do género masculino e 17 do sexo feminino; média das idades=15,19). O GCC foi dividido em dois sub-grupos, o sub-grupo TF (n=15, 8 do género masculino e 7 do sexo feminino; média das idades=15,00) e o sub-grupo CIV (n=17, 7 do género masculino e 10 do sexo feminino; média das idades=15,35). Foram recolhidos os dados clínicos e demográficos mais relevantes e, num único momento temporal, foi aplicado um conjunto de instrumentos neuropsicológicos: FCR (cópia e memória); PC da bateria *Behavioural Assessment of the Dysexecutive Syndrome* (BADS); o StroopP, StroopC e StroopI, Cd (parte B) e MD (directo e inverso), da Escala de Inteligência de Wechsler para Crianças - III *(*WISC-III); o TMT (parte A e B) e a sub-prova ML da Escala de Memória de Wechsler-III (WMS-III).

Resultados: não foram encontradas diferenças entre os grupos, no que respeita às variáveis demográficas. Quando realizada a comparação dos resultados obtidos nas provas neuropsicológicas pelos GC e GCC, verificamos que existem diferenças estatisticamente significativas entre os dois grupos ($p \leq 0{,}001$), excepto na prova Cd ($p=0{,}654$) e ML ($p=0{,}095$). Entre os sub-grupos CIV e TF, verificamos que existem diferenças estatisticamente significativas no desempenho neuropsicológico nas provas DD ($p=0{,}049$), StroopC ($p=0{,}018$), StroopI ($p=0{,}024$) e TMT-B (p=0,049). No estudo correlacional efectuado entre as variáveis neo-natais e o desempenho nas provas neuropsicológicas, verificamos que no GCC e nos sub-groupos CIV e TF, existem correlações positivas e negativas entre as variáveis neonatais e os domínios neurocognitivos avaliados.

Conclusão: Os adolescentes com CC apresentam piores resultados nas provas neuropsicológicas, quando comparados com o GC. Foram encontradas diferenças entre os sub-grupos cianótico e acianótico. Verificamos também que existe uma forte influência das variáveis neonatais no desenvolvimento dos vários domínios neurocognitivos estudados.

Palavras - Chave: CC, neurodesenvolvimento, variáveis neonatais.

Abstract

Purpose: Evaluate the neurocognitive performance in adolescents with CHD and understand if the neonatal variables (Apgar scores; head circumference, the length and weight), are related with the neurocognitive performance of the adolescents mentioned above.

Procedure: 48 adolescents have participated, 26 male and 22 female, aged between 13 and 18. The sampling was divided In two groups, The GC (n=16, 11 male and 5 female; average age = 15,69) and the GCC (n=32, 15 male and 17 female, average age = 15,19).O GCC was still divided in 2 sub groups, the sub group TF (n=15; 8 male and 7 female; average age = 15,00) and the subgroup CIV(n=17; 7 male and 10 female; average age = 15,35). The most relevant clinic and demographic date were collected and on a single moment in time, a group of neuropsychological instruments were applied: FCR (copy and memory); Key Search from *Behavioural Assessment of the Dysexecutive Syndrome* (BADS); the StroopW, StroopC e StroopI, Cd (parte B) e MD (directo e inverso), da Escala de Inteligência de Wechsler para Crianças - III *(*WISC-III); o TMT (parte A e B) e a sub-prova ML da Escala de Memória de Wechsler-III (WMS-III).

Results: differences between groups were not found in what concerns to demographic variables. When comparing the obtained results on the neuropsychological tasks by GC and GCC, we verified that there are significant statistically differences between the two groups, in what concerns the performance in all the neuropsychological tasks ($p \leq 0{,}001$), unless in proof Cd (p=0,654) and ML(p=0,095). Between the CIV and TF sub groups, we verified that there are significant statistically differences between the two groups, in what concerns the neuropsychological performance in DD (p=0,049), StroopC (p=0,018), StroopI (p=0,024) e TMT-B (p=0,049). In the correlational study between the neonatal variables and performance in the neuropsicological tasks, we verified that in GCC, sub-group CIV and sub-group TF, there are positive and negative correlations between neonatal variables and the neurocognitive domains evaluated.

Conclusions: Adolescents with CHD show worst neuropsychological performance, when compared with the control group. Diferences were found between cyanotic and acyanotic groups. A great influence from neonatal variables was found in neurocognitive domains' progress.

Key Words: CHD, neurodevelopment, neonatal variables.

Lista de Abreviaturas

GC – Grupo de Controlo

GCC – Grupo Cardiopatias Congénitas

CIV – Grupo CIV

TF – Grupo TF

DD – Dígitos Directos

DI – Dígitos Inversos

PC – Procura da Chave

ML – Memória Lógica

Cd - Código

FCRc – Figura Complexa de Rey (cópia)

FCRm – Figura Complexa de Rey (memória)

StroopP – Stroop Palavras

StroopC – Stroop Cores

StroopI – Stroop Interferencia

TMT – A – Trail Making Test parte A

TMT- B – Trail Making Test parte

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Capítulo I - Introdução

As Cardiopatias Congénitas (CC) representam a mais comum causa de morbilidade e mortalidade infantil (Donofrio & Massaro, 2010).

Na década de 50, apenas 20% das crianças que nasciam com CC moderada ou severa sobreviviam ao primeiro ano de vida. Em consequência da evolução nos tratamentos e cirurgias, nos dias de hoje, 90% das crianças que nascem com CC sobrevivem até à idade adulta (Reid, Webb, Barzel, McCrindle, Irvine & Siu, 2006).

As recentes melhorias em técnicas de intervenção cirúrgica permitiram a paliação ou correcção precoce de CC complexa na infância, resultando assim numa redução da taxa de mortalidade. Por conseguinte, a expectativa de vida destes doentes melhorou favoravelmente (Brickner, Hills & Lange, 2000b).

Neste sentido, as melhorias no prognóstico e tratamento de cardiopatias congénitas, provocaram uma diminuição de 30% na mortalidade destes doentes (Nousi & Christou, 2010).

A condição crónica da CC e o crescimento desta população leva a que os problemas médicos, psicológicos e comportamentais sejam de extrema importância para os profissionais de saúde. Assim, o principal foco de atenção não se direcciona apenas para o alcance da sobrevivência a longo prazo, mas sim para a compreensão dos aspectos relacionados com a qualidade de vida, ajustamento psicossocial, morbilidade psiquiátrica e desenvolvimento neurocognitivo desta população (Massaro, El-dib, Glass & Aly, 2008).

Embora a maioria das crianças com CC sobreviva nos dias de hoje, mais de metade apresentará um desenvolvimento neurológico comprometido em vários domínios (Bellinger, Wypij, Duplessis, Rappaport, Jonas, Wernovsky & Newburger, 2003b). Neste sentido, torna-se fundamental verificar o impacto das cardiopatias congénitas no desenvolvimento cerebral do feto e da criança, bem como as alterações do crescimento fetal intra-uterino (Denofrio & Massaro, 2010), tendo em conta as repercussões a longo prazo no desenvolvimento cognitivo destas crianças.

Assim foi criada uma linha de Investigação intitulada de "Impacto do Desenvolvimento Fetal no Funcionamento neurocognitivo em Adolescentes com

Cardiopatias Congénitas", cujo objectivo é o estudo do desempenho neurocognitivo em adolescentes com Cardiopatias Congénitas Cianóticas e Acianóticas (TF e CIV, respectivamente) e compreender se variáveis neonatais (Índice de Apgar, o perímetro cefálico, o comprimento e peso), estão relacionadas com o desempenho neurocognitivo dos adolescentes supra mencionados. Para o efeito, foi seleccionado um dos hospitais com maior representatividade na zona norte do país – o Hospital de São João, bem como escolas do grande Porto.

Com a finalidade de atingir os objectivos propostos, foi seleccionada uma amostra, constituída por 48 adolescentes, do género feminino e masculino, com idades compreendidas entre os 13 e os 18 anos de idade. A mesma foi dividida em dois grupos, o Grupo de Controlo e o Grupo de Cardiopatias Congénitas. Este último era constituído por dois sub-grupos, TF e CIV. Os adolescentes do primeiro grupo frequentavam escolas do distrito do Porto e os adolescentes do grupo de Cardiopatias Congénitas estavam integrados na consulta externa de Cardiologia Pediátrica.

Todos os elementos da amostra foram submetidos a uma avaliação neuropsicológica, constituída por um conjunto de instrumentos que permitia avaliar o maior número de funções cognitivas, no menor tempo possível. As informações adicionais e necessárias, referentes ao grupo de Cardiopatias Congénitas foram consultadas nos arquivos dos processos clínicos, com a colaboração da equipa administrativa do serviço em questão.

Estruturalmente, a dissertação encontra-se dividida em vários capítulos. O primeiro e segundo capítulos, dizem respeito a uma breve introdução ao tema, propósitos da investigação e ao racional teórico, onde é realizada a conceptualização do tema, respectivamente. O terceiro capítulo é referente à caracterização do estudo, onde é exposto o objectivo central da investigação, bem como os objectivos específicos, as hipóteses formuladas e as variáveis em estudo. Já o quarto capítulo diz respeito à metodologia, onde é efectuada uma caracterização da amostra e onde são apresentados os instrumentos de avaliação, o procedimento metodológico, o desenho e o procedimento estatístico. No quinto capítulo são apresentados os resultados, para posteriormente no sexto capítulo serem integrados com a componente teórica. Por último, são apresentadas as conclusões e considerações finais que surgiram após a realização do estudo.

Capítulo II – Racional Teórico

Cardiopatias Congénitas

As Cardiopatias Congénitas podem ser descritas como um grave defeito estrutural do coração ou dos grandes vasos, que afecta potencial e significativamente o seu funcionamento. È uma doença cronica, normalmente diagnosticada ainda no período fetal (Nousi & Christou, 2010). Assim, as malformações cardiovasculares congénitas são consequência de um desenvolvimento embrionário anómalo de uma estrutura normal, ou da incapacidade de tal estrutura prosseguir para além de um estágio inicial de desenvolvimento fetal ou embrionário (Kasper *et al*., 2006,p.1448).

Em cerca de 80% a 90% dos casos, a origem das cardiopatias congénitas não é reconhecida. No entanto, acredita-se ser multifactorial, resultante da combinação da predisposição genética e de factores ambientais (Gandhi & Sreekantam, 2010).

Alguns pacientes com cardiopatias congénitas apresentam apenas defeitos leves, com relativamente pouca necessidade de cuidados médicos, enquanto outros, com defeitos mais severos, requerem serviços médicos frequentes (Hoffman & Kaplan, 2002).

Neste sentido, as cardiopatias congénitas podem ser classificadas em 3 grupos de acordo com a sua gravidade, como ligeiras, moderadas e graves (Miatton, De Wolf, François, Thiery & Vingerhoets, 2006).

As cardiopatias congénitas ligeiras são caracterizadas por subtis limitações da vida do paciente, que geralmente não revela sintomas e que na maior parte dos casos não necessita de intervenção cirúrgica, devido à resolução espontânea da lesão (Miatton *et al*., 2006).

Os pacientes com cardiopatias congénitas moderadas tendem a necessitar de intervenções cirúrgicas, que assumem um papel preponderante na sua evolução e expectativas de vida (Kovacs, Sears & Saidi, 2005).

Por ultimo, as cardiopatias congénitas complexas ou graves incluem a maioria dos pacientes que se apresentam gravemente doentes no período neonatal ou na primeira infância. Desta forma são necessários cuidados especializados de monitorização

cardíaca frequente. Os pacientes com CC grave são sujeitos a uma/múltiplas intervenções cirúrgicas (Miatton *et al.*, 2006).

A incidência de cardiopatias congénitas na população Portuguesa é de 5 a 12 mil recém - nascidos. Entre as malformações congénitas, é a patologia mais frequente, sendo que nos últimos anos, houve um decrescimento de óbitos no primeiro ano de vida, de 11.5% em 2000 para 8.7% em 2004 (Direcção Geral de Saúde, 2006).

Cardiopatias Congénitas Cianóticas e Acianóticas

As cardiopatias congénitas podem ser organizadas clinicamente em dois grandes grupos: as cardiopatias congénitas cianóticas e cardiopatias congénitas acianóticas. Esta distinção é classificada de acordo com o grau de saturação de oxigénio no sangue.

✓ *As Cardiopatias Congénitas Cianóticas*

A cianose pode ser identificada pela coloração azulada da pele, consequência de um baixo nível de oxigénio no sangue. Assim, os pacientes com cardiopatias congénitas cianóticas padecem de uma dessaturação do oxigénio arterial causado por um desvio do sangue venoso sistémico para a circulação arterial (Brickner, Hillis & Lange, 2000b). Nestes casos, a maioria das crianças com cardiopatia cianótica necessitam de intervenção cardíaca para sobreviverem até à idade adulta. Nesta fase da vida, as causas de cardiopatia cianótica mais frequentes são a Tetralogia de Fallot (TF) e o Síndrome de Eisenmenger (Brickner *et al*., 2000b). No entanto, outras patologias estão inseridas nas cardiopatias cianóticas, como a Transposição das Grandes Artérias (TGA), o Coração Univentricular e a Anomalia de Ebstein (AE) (Brickner *et al*., 2000b).

A Tetralogia de Fallot é caracterizada por um grande defeito do septo ventricular, aorta cavalgada sobre a comunicação interventricular, obstrução do fluxo do ventrículo direito e hipertrofia do ventrículo direito (Brickner *et al*., 2000b).

Durante a infância são identificados episódios de taquipnéia e hiperpnéia nos pacientes com TF, provocando um agravamento da cianose, verificando-se em alguns casos perda de consciência, convulsões, acidentes vasculares cerebrais ou até mesmo a morte (Brickner *et al*., 2000b).

Na idade adulta, os indivíduos tendem a apresentar dispneia e dificuldades no que concerne à prática de exercício físico. Os problemas de cianose crónica podem levar à ocorrência de um acidente vascular cerebral.

O Síndrome de Eisenmenger é caracterizado por amplas comunicações entre as duas circulações no nível aorticopulmonar, ventricular ou auricular e shunts bidireccionais ou predominantemente direita-esquerda devidos à resistência elevada e hipertensão pulmonar obstrutival. Pacientes que apresentem este síndrome, podem manifestar dispneia e baixa tolerância á prática do exercício físico, palpitações, arritmias e risco de morte súbita (Brickner *et al.*, 2000b).

A Transposição das Grandes Artérias é caracterizada pela aorta que surge numa posição anterior desde o ventrículo direito e a artéria pulmonar que surge do ventrículo esquerdo, ocorrendo assim uma separação integral da circulação pulmonar e sistémica (Brickner, *et al.*, 2000b). Os pacientes com TGA apresentam cianose e taquipneia desde o momento em que nascem, necessitando de intervenção cirúrgica para que haja um prognóstico favorável (Brickner *et al.*, 2000b).

O Coração Univentricular é caracterizado por uma abertura para a câmara ventricular única das duas válvulas atrioventriculares ou de uma válvula atrioventricular. Um fluxo sanguíneo pulmonar normal, a resistência pulmonar e a correcta função ventricular são fundamentais para a sobrevivência até à idade adulta de pacientes com este tipo de cardiopatia (Kasper *et al*., 2006).

A Anomalia de Ebstein é representada por uma anomalia da válvula tricúspide, consequência de uma fixação anormal das cúspides valvulares. Quando a anomalia é diagnosticada durante a vida fetal, a taxa de mortalidade intrauterina é elevada. Pacientes com anomalia de Ebstein apresentam insuficiência cardíaca grave, durante a vida fetal e arritmia supraventricular na adolescência e idade adulta. Os pacientes que apresentam a patologia e comunicação interauricular estão em risco de embolia cerebral e morte súbita (Brickner *et al*., 2000b).

✓ *As Cardiopatias Congénitas Acianóticas*

Ao contrário das cardiopatias cianóticas, as cardiopatias congénitas acianóticas são caracterizadas por uma saturação do oxigénio no sangue arterial e por uma coloração normal da pele (Nousi & Christou, 2010).

No grupo das cardiopatias congénitas acianóticas podemos descrever o Defeito do Septo Ventricular, o Defeito do Septo Auricular, a Estenose da Aorta, a Estenose Pulmonar e a Coartação da Aorta.

No que respeita ao Defeito do Septo Ventricular, inúmeras consequências fisiológicas estão associadas, nomeadamente o tamanho do defeito e a resistência à circulação vascular sistémica e pulmonar. Anatomicamente está localizado em 70% dos casos na porção membranosa do septo interventricular, em 20% na porção na porção muscular do septo, 5% abaixo da válvula aórtica e 5% próximo das válvulas mitral e tricúspide (Brickner, Hillis & Lange, 2000a).

O defeito do septo ventricular é a CC mais comum nas crianças, ocorrendo em frequência semelhante nos rapazes e nas raparigas.

Os adultos com um defeito pequeno manifestam tensão arterial normal e são geralmente assintomáticos. No entanto, quando o defeito é de tamanho significativo, os pacientes que conseguem chegar à idade adulta sofrem de insuficiência ventricular esquerda, hipertensão pulmonar ou falha no ventrículo direito (Brickner, *et al.*, 2000a).

O Defeito do Septo Auricular é caracterizado pela comunicação persistente entre ambos os átrios, ocorrendo o desvio do sangue de um átrio para o outro (Brickner, *et al*, 2000a). Do ponto de vista anatómico, pode tomar a forma de ostium primum na zona inferior do septo auricular, ostium secundum ao nível da fossa ovalis, ou sinus venosus na zona do septo auricular superior (Brickner, *et al.*, 2000a).

A Estenose da Aorta é uma das cardiopatias congénitas mais frequentes. É caracterizada pela estenose congénita da válvula aórtica, estenose subaórtica leve, estenose aórtica supravalvar e miocardiopatia obstrutiva hipertrófica, cujas malformações obstruem a saída do ventrículo esquerdo (Kasper *et al*., 2006). Na idade adulta, os pacientes com diagnóstico assintomático, poderão ter uma vida normal. No entanto, nos pacientes que revelam sintomas é recomendada a cirurgia de substituição da válvula. A sintomatologia mais comum desta patologia é a angina de peito, sincope e

insuficiência cardíaca. A incidência da estenose da aorta é superior no sexo masculino. (Brickner, *et al*., 2000a).

A Estenose pulmonar pode ser descrita como uma obstrução do fluxo de saída do ventrículo direito, podendo ocorrer ao nível valvular, supravalvular ou subvalvular, ou simultaneamente nestes níveis (Brickner, *et al*., 2000a). Normalmente os pacientes com esta patologia são assintomáticos, no entanto, em casos de maior gravidade é possível descrever sintomas como dores no peito, dispneia, fadiga em situações de esforço e insuficiência do ventrículo direito (Kasper *et al.*, 2006).

A Coartação da Aorta pode ser definida como um estreitamento da luz da aorta, passível de aparecer em qualquer segmento ao longo do seu comprimento, sendo mais frequentemente localizado no segmento distal à origem da artéria subclávia esquerda, próximo da inserção do ligamento arterial (Brickner *et al*., 2000a).

Nos pacientes com este tipo de CC, a sintomatologia mais frequente é a hipertensão arterial, cefaleias, tonturas, palpitações e insuficiência cardíaca (Brickner, *et al.*, 2000a).

Cardiopatias Congénitas e Desenvolvimento Fetal

Devido ao aumento da sobrevivência de pacientes com cardiopatia congénita, um número considerável de estudos têm sido realizados com a finalidade de verificar o impacto da mesma no funcionamento psicológico e cognitivo em crianças e adolescentes (Karsdorp, Everaerd, kindt & Mulder, 2007). Neste sentido, o objectivo da assistência médica não se direcciona apenas para a sobrevivência, mas também para as dificuldades que estes pacientes enfrentam ao nível psicológico e cognitivo (Karsdorp, *et al.*, 2007)

Vários estudos têm identificado um desenvolvimento neurológico anormal em 25% da população com cardiopatias congénitas (Bellinger, Wypij, Kuban, Rappaport, Hickey, Wernovsky, Jonas, & Newburger, 1999). A identificação precoce dos défices desenvolvimentais e intervenções cirúrgicas corretivas posteriores são de extrema importância para garantir o desenvolvimento ideal nestas crianças (Miatton, De Wolf, François, Thiery & Vingerhoets, 2006).

Muitas crianças que foram submetidas a cirurgia cardíaca no período neonatal, demonstraram um padrão de desenvolvimento de problemas neurológicos, caracterizados por défices cognitivos, de atenção e funcionamento executivo, de capacidades visuo-motoras e visuo-espaciais, de atraso na aquisição da linguagem expressiva e de dificuldades de aprendizagem (Bellinger *et al.*, 1999; Miatton, De Wolf, François, Thiery & Vingerhoets, 2007a; Daliento, Mapelli & Volpel, 2006; Brosig, Mussatto, Kuhn & Tweddell, 2007; Gerdes & Flynn, 2010; Fuller, Nord, Gerdes, Wernovsky, Jarvik, Bernbaum, Zackai & Gaynor, 2009).

Tradicionalmente, os estudos realizados acerca dos resultados neurológicos em crianças com cardiopatias congénitas, têm-se focado em factores relacionados com a cirurgia, nomeadamente quando a perfusão cerebral pode estar comprometida durante a circulação extra corporal (Donofrio & Massaro, 2010). No entanto, outros estudos têm demostrado uma alta prevalência de anormalidades neurológicas anatómicas e funcionais antes da cirurgia em recém-nascidos com cardiopatia congénita (Limperopoulos, Tworetzky, McElhinney, Newburger, Brown, *et al.*, 2010; Miller, McQuillen, Hamrick, Xu, Glidden *et al.*, 2007; Watanabe, Matsui, Matsuzawa, Tanaka, Noguchi, *et al.*, 2009), bem como uma relação entre estas anormalidades e o desenvolvimento cognitivo durante a infância.

Segundo alguns estudos, foram encontradas anormalidades neurocomportamentais anteriores à cirurgia em mais de 50% dos recém-nascidos (menos de um mês) e 38% de crianças (entre 1 mês e 2 anos de idade) com cardiopatias congénitas. Estas anormalidades incluem hipotonia, agitação/inquietação, assimetria motora, letargia e características autistas. Geralmente estas anormalidades persistem ou pioram no período pós-operatório, onde surgem também anormalidades nos nervos cranianos (Limperopoulos, Majnemer, Shevell, Rosenblatt, Rohlicek, & Tchervenkov, 1999; Limperopoulos, Majnemer, Shevell, Rosenblatt, Rohlicek, & Tchervenkov, 2000). Ainda no período pré operatório, outras anomalias podem surgir. As mais comuns podem ser caracterizadas por agenesia ou disgenesia do corpo caloso, holoprosencefalia, microcefalia, lisencefalia (Chock, Reddy, Bernstein & Madan, 2006), abertura do opérculo, enfartes isquémicos e lesão da substancia branca caracterizada por leucomalácia periventricular (PVL) (Mahle, Tvani, Zimmerman, *et al.*, 2002). Os recém - nascidos com cardiopatia congénita apresentam potenciais risco de apresentarem hipoxia, hipotensão e acidose.

Este facto revela a existência de uma forte possibilidade de que as malformações cardíacas congénitas e as anomalias no crescimento fetal estão causalmente relacionadas (Rosenthal, 1996; Watanabe *et al.*, 2009), sendo suportada também a hipótese de que as anormalidades do desenvolvimento cerebral, as alterações hemodinâmicas intra-uterinas e as anormalidades cerebrais congénitas e lesões cerebrais adquiridas, relacionadas com cianose prolongada e hipoperfusão após o nascimento (Donofrio & Massaro, 2010), contribuem para efeitos adversos no desenvolvimento neurológico em crianças com cardiopatia congénita (Limperopoulos, Tworetzky, McElhinney, Newburger, Brown, *et al*., 2010)

Os recém-nascidos com cardiopatias congénitas apresentam uma alta frequência de lesão cerebral adquirida, comprovada em estudos de ressonância magnética no período peri-operatório. A elevada incidência de lesão da substancia branca em recém-nascidos, sugere uma vulnerabilidade única que pode estar relacionada com um atraso no desenvolvimento do cérebro. Estas anormalidades no desenvolvimento cerebral em recém-nascidos com cardiopatias congénitas podem reflectir anormalidades no fluxo sanguíneo cerebral do feto (McQuilen & Miller, 2010).

Para muitos recém-nascidos, a doença congénita do coração parece ser uma anomalia isolada, presumindo-se que o cérebro tem um potencial de desenvolvimento normal (Shillingford, Glanzman, Ittenbach, Clancy, Gaynor & Wernovsky, 2007). No entanto, o desenvolvimento do cérebro e do coração ocorrem simultaneamente no feto com cardiopatia congénita, sendo que os primeiros programas morfogenéticos de cada órgão, compartilham vias genéticas em comum.

Enquanto que o coração é morfologicamente maduro por volta da sétima semana de gestação, o desenvolvimento cerebral estende-se ao longo de um período de tempo mais alargado, com distintos eventos morfológicos que ocorrem nos primeiros dois trimestres (McQuillen, Goff & Licht, 2010), nomeadamente a migração e arborização neuronal, sinaptogénese, morte celular programada, maturação dos oligodendrócitos e extensa reorganização das conexões sinápticas (Limperopoulos, Tworetzky, McElhinney, Newburger, Brown *et al.*, 2010). Esta fase é seguida de um período prolongado de crescimento cerebral marcante e dependente da formação e aperfeiçoamento de conexões no terceiro trimestre e período pós-natal (McQuillen, Goff & Licht, 2010). Este desenvolvimento está associado com o aumento da actividade metabólica, em que o cérebro está dependente do coração para o fornecimento de oxigénio e de nutrientes.

Contudo, a presença de cardiopatias congénitas leva a anormalidades no fluxo sanguíneo fetal, que consequentemente gera um crescimento e desenvolvimento cerebral comprometido, devido às relações complexas entre linhagens de células comuns, programação genética, consequências fisiológicas da alteração do fluxo sanguíneo cerebral e à dinâmica da distribuição de oxigénio no cérebro em desenvolvimento (McQuilen & Miller, 2010). Neste sentido, existe uma boa evidencia que estes factores assumem um papel preponderante no crescimento e maturação cerebral (DiNardo, 2010).

Na circulação fetal normal, as trocas gasosas ocorrem através da placenta. O sangue desoxigenado vindo da veia cava superior, entra directamente no ventrículo direito e através do canal arterial para a placenta. A válvula de Eustáquio e o septo arterial movem-se em conjunto para dirigir o sangue venoso da veia cava inferior hepática para o ventrículo direito e o sangue oxigenado do canal venoso sobre o forâmen oval através do ventrículo esquerdo para a aorta e circulação cerebral.

Em situações em que a oxigenação fetal está comprometida, há uma redistribuição sanguínea para a circulação cerebral, fenómeno conhecido como "brain sparing" (Donofrio & Massaro, 2010), resultando num padrão de restrição global do crescimento somático, com relativa preservação do crescimento da cabeça. (McQuilen & Miller, 2010). Este fenómeno hemodinâmico é representado pelo aumento do fluxo diastólico nas artérias cerebrais e pela diminuição do fluxo diastólico nas artérias aorta descendente e umbilical (Donofrio & Massaro, 2010). No entanto, regiões específicas do cérebro, poderão estar mais protegidas do que outras. Segundo um estudo de Dubiel, Gunnarsson & Gunnarsson (2002), realizado em gravidas com gestações complicadas devido a hipertensão materna e disfunção da placenta, a vasodilatação cerebral foi encontrada em 41% na artéria cerebral anterior, em 30% na artéria cerebral posterior e em 24% na artéria cerebral média. Assim, as artérias cerebrais anteriores apresentam uma melhor resposta autorreguladora, sendo que a redistribuição do fluxo sanguíneo favorece a perfusão dos lobos frontais. No entanto, as artérias cerebrais médias apresentam-se como menos reactivas (Donofrio & Massaro, 2010).

Paradoxalmente, este mecanismo tem sido descoberto como um percursor de um desenvolvimento neurológico adverso, pois uma vez que a vasodilatação cerebral ocorre quando a oxigenação fetal está comprometida, este mecanismo de protecção pode ser insuficiente para manter o crescimento e desenvolvimento cerebral normal em situações de stress prolongado *in útero* (Donofrio & Massaro, 2010).

Na circulação sanguínea fetal normal, o sangue oxigenado é dirigido para o cérebro e o sangue desoxigenado para a placenta (Donofrio & Massaro, 2010).

Como referido anteriormente, estudos demonstram que a existência de anormalidades na circulação sanguínea, como ocorre em cardiopatias complexas, nomeadamente no Síndrome Hipoplásico do Coração Esquerdo, Transposição Das Grandes Artérias, Tetralogia de Fallot, podem contribuir para um desenvolvimento cerebral anormal (Limperopoulos, Tworetzky, McElhinney, Newburger, Brown *et al*., 2010; Donofrio & Massaro, 2010;McQuillen & Miller, 2010).

Em fetos com Sindrome Hipoplasico do Coração Esquerdo existe provavelmente um aumento da resistência ao fluxo cerebral, em que a circulação sanguínea retrocede através de uma hipoplasia istmo-aórtica para chegar ao cérebro; fetos com Transposição da Grandes Artérias, o sangue venoso da circulação cerebral volta directamente para o cérebro; fetos com Tetralogia de Fallot e Sindrome Hipoplasico do Coração Direito, o sangue relativamente desoxigenado entra na circulação cerebral devido à mistura intracardíaca.

Assim, o tipo de lesão afecta não apenas a origem do fluxo sanguíneo cerebral, mas também o grau de sangue desoxigenado distribuído através da circulação cerebral (Donofrio & Massaro, 2010).

Todas estas alterações na circulação sanguínea poderão ser percursores de um crescimento somático anormal in – útero, influenciando a antropometria neonatal (Rosenthal, 1996).

Comparativamente com recém – nascidos sem patologia congénita cardíaca, recém - nascidos com Síndrome Hipoplásico do Coração Esquerdo, apresentam peso, comprimento e perímetro cefálico menores do que o normal e o volume da cabeça é desproporcionalmente pequeno relativamente ao peso. Os recém-nascidos com Transposição das Grandes Artérias, apresentam peso normal, mas um menor perímetro cefálico relativamente ao peso. Por outro lado, os recém-nascidos com Tetralogia de Fallot têm proporções normais, no entanto, apresentam peso, comprimento e perímetro cefálico menores do que o normal. As crianças com Coartação da Aorta, apresentam um maior volume da cabeça, relativamente ao peso ao nascimento (Donofrio & Massaro, 2010; McQuilen & Miller, 2010; Hinton, Andelfinger, Sekar *et al*, 2008; Rosenthal, 1996).

Muitos factores podem contribuir para um baixo peso ao nascimento, incluindo síndromes genéticos, insuficiência placentária, restrição do crescimento intra-uterino. Todos eles podem aumentar o risco de atrasos no neurodesenvolvimento.

Recentemente, Gaynor, Jarvik, Bernbaum, Gerdes, Wernovsky *et al*., (2006), mostraram, que no geral, os factores inerentes ao paciente, como peso à nascença, perímetro cefálico ao nascer e índices de Apgar no 1º e no 5º minuto, explicam melhor a vulnerabilidade do índice de desenvolvimento mental e psicomotor, do que os factores intra-operatórios (peso na cirurgia, tempo de resfriamento, paragem circulatória por hipotermia profunda, duração da circulação extracorporal, baixa temperatura nasofaríngea).

Cardiopatias Congénitas e Desenvolvimento Cognitivo

Como mencionado anteriormente, a alteração da circulação sanguínea fetal subjacente à presença de CC, assume uma influência significativa no desenvolvimento cerebral e crescimento somático intra-uterino. Neste sentido, estes factores, revelam-se como possíveis percursores de anormalidades no desenvolvimento neurocognitivo a longo prazo.

Diferentes estudos apontam para uma normalidade do índice do QI, em crianças com CC, comparativamente a crianças saudáveis. Assim, a avaliação cognitiva não deve ser restrita apenas a esta medida, mas também devem ser realizadas avaliações a outros domínios, nomeadamente a aprendizagem, a memória, a velocidade psicomotora, atenção, concentração, raciocínio, cálculo e funções executivas (Daliento *et al*., 2006)

Vários estudos têm referido a existência de alterações neurodesenvolvimentais em crianças com cardiopatias congénitas (Brosig, Mussatto, Kuhn & Tweddell, 2007; Bellinger & Newburger, 2010; Gerdes & Flynn, 2010). No entanto, em muitas investigações, as avaliações são realizadas em crianças, maioritariamente na primeira infância, não permitindo a avaliação de determinadas funções que têm maturação mais tardia, nomeadamente, o funcionamento executivo (Bellinger & Newburger, 2010). Assim, os défices neurocognitivos nas crianças com Cardiopatias Congénitas são reconhecidos mais tarde, sobretudo em idade escolar (Gerdes & Flynn, 2010).

Segundo Miatton e colaboradores (2008), as crianças com CC, em idade escolar, apresentam alterações ao nível do funcionamento sensoriomotor, funcionamento executivo, atenção, memória e linguagem, quando comparadas a crianças saudáveis. Neste estudo, foi também analisada a percepção dos pais em relação às dificuldades dos filhos, onde foram percepcionadas dificuldades em vários domínios, nomeadamente na atenção sustentada e atenção dividida, na memória e na capacidade de aprendizagem.

No que respeita domínio atencional, foram evidenciadas alterações nas crianças com Cardiopatias Congénitas (Bellinger, 2010; Gerdes & Flynn, 2010). Num estudo realizado com 109 crianças, submetidas a cirurgia corretiva no período neonatal, verificou-se que uma percentagem significativa apresentava défice de atenção e hiperatividade, na idade escolar. Neste estudo, foram atribuídas pontuações de "alto risco", por 30% dos pais, para défice atencional. (Shillingford, *et a*l., 2008).

Outras pesquisas apontam também alterações, sobretudo ao nível das funções executivas, velocidade de processamento da informação, capacidades visuo-espaciais, funções motoras e linguagem (Daliento, Mapelli, Russo, Scarso, Limongi, *et al*., 2005; Bellinger & Newburger, 2010; Gerdes & Flynn, 2010).

Vários estudos mencionam que crianças com CC apresentam alterações no funcionamento executivo (Bellinger *et al*., 2003b; Miatton *et al*., 2007a; Miatton *et al*., 2007b). Este constructo abrange capacidades de ordem cognitiva superior, que estão relacionadas com o planeamento estratégico, a flexibilidade cognitiva, a auto-regulação e o comportamento objectivo directo (Daliento, Mapelli & Volpe, 2006). O funcionamento executivo está relacionado com a integridade do córtex frontal e pré-frontal, que em caso de lesão, leva a dificuldades de julgamento, planeamento, tomada de decisão, resolução de problemas e desinibição comportamental (Royall, Lauterbach, Cummings, Reeve, Rummans, *et al*., 2002).

Quando comparado o desempenho cognitivo em crianças com CC cianótico e acianótico, alguns estudos demonstram não existirem diferenças significativas entre ambos (Oates, Simpson , Cartmill & Turnbull, 1995; Hövels-Gürich, *et al.*, 2006; Miatton *et al.*, 2007a; Miatton *et al.*, 2007b). No entanto, as crianças que tiveram hipoxemia na fase pré-operatória, devido ao defeito cardíaco cianótico, evidenciam um maior risco para apresentar disfunção ao nível da atenção, no domínio do controlo executivo, em comparação com crianças com CC acianóticas (Hövels-Gürich, et al., 2007).

Capítulo III - Caracterização do Estudo

Objectivos

O presente trabalho de investigação assume como objectivo estudar o desempenho neurocognitivo em adolescentes com Cardiopatias Congénitas Cianóticas e Acianóticas (TF e CIV, respectivamente).

Apresenta ainda como finalidade, compreender se variáveis neonatais (Índice de Apgar, o perímetro cefálico, o comprimento e peso), estão relacionadas com o desempenho neurocognitivo dos adolescentes supra mencionados.

Desta forma, foram estabelecidos os seguintes objectivos específicos:

1. Comparar o desempenho neurocognitivo em adolescentes com Cardiopatias Congénitas;

2. Estabelecer comparação do desempenho neurocognitivo em adolescentes com Cardiopatias Congénitas Cianóticas (TF) e Acianóticas (CIV);

3. Verificar a existência de uma correlação entre variáveis neonatais e desempenho neurocognitivo em adolescentes com Cardiopatias Congénitas;

4. Averiguar a existência de uma relação entre as variáveis neonatais nas Cardiopatias Congénitas Cianótica e Acianótica.

O estudo efectuado torna-se relevante na medida em que permite uma caracterização ao nível cognitivo de adolescentes com CC, de modo a compreender quais os défices cognitivos mais comummente presentes em dois subgrupos específicos, nomeadamente, no subgrupo CIV (cardiopatia acianótica) e no sub-grupo TF (cardiopatia cianótica). A pertinência da investigação é ainda realçada pelo estudo correlacional efectuado entre as variáveis neonatais e o desempenho neuropsicológico, Todo o trabalho desenvolvido revela-se extremamente importante na medida em que poderá ser útil para posteriormente implementar programas de estimulação, para assim

intervir precocemente nas crianças com este tipo de patologia, de modo a minimizar o impacto da mesma em suas vidas.

Hipóteses

Para o presente estudo foram desenvolvidas as seguintes hipóteses:

Hipótese I – Os adolescentes com CC apresentam um pior desempenho nas provas neuropsicológicas, comparativamente com o grupo de adolescentes saudáveis;

Hipótese II – O sub-grupo TF (cardiopatia cianótica) demonstra uma pior performance nas provas neuropsicológicas, quando comparado com o sub-grupo CIV (cardiopatia acianótica);

Hipótese III – Existem correlações entre as variáveis neonatais e o desempenho nas provas neuropsicológicas no GCC;

Hipótese IV - Existem correlações entre as variáveis neonatais e o desempenho nas provas neuropsicológicas no sub-grupo CIV (cardiopatia acianótica);

Hipótese V - Existem correlações entre as variáveis neonatais e o desempenho nas provas neuropsicológicas no sub-grupo TF (cardiopatia cianótica).

Capitulo IV - Metodologia

Caracterização da Amostra

A amostra em estudo é constituída por 48 adolescentes, do género feminino e masculino, com idades compreendidas entre os 13 e os 18 anos de idade. A mesma foi dividida em dois grupos, o Grupo de Controlo e o Grupo de doentes com Cardiopatias Congénitas. Este último foi ainda dividido em dois sub-grupos, o sub-grupo de doentes com TF e o sub-grupo de doentes com Defeito do septo Interventricular.

A amostra foi designada considerando alguns critérios de exclusão, nomeadamente a presença de patologia psiquiátrica, neurológica ou outra perturbação sistémica susceptível de interferir com o funcionamento neurocognitivo.

As tabelas 1 e 2, seguidamente expostas apresentam a caracterização dos grupos, no que concerne às variáveis demográficas (idade, género e anos de escolaridade).

Tabela 1

Caracterização dos grupos da amostra considerando as variáveis demográficas (idade, género e anos de escolaridade).

	GC			GCC		
	N=16			N=32		
	M	DP	n	M	DP	n
Idade	15.69	1.45	-	15.19	1.82	-
Género (M/F)	-	-	11/5	-	-	15/17
Anos de Escolaridade	10.13	1.36	-	9.06	2.21	

No que respeita à idade (p=.344), ao género (χ^2= 2,056; p=.152) e aos anos de escolaridade (p=.086), não existem diferenças estatisticamente significativas entre os dois grupos.

Tabela 2

Caracterização dos sub-grupos da amostra, no que respeita às variáveis demográficas.

	CIV N=17			TF N=15		
	M	DP	n	M	DP	n
Idade	15.35	1.66	-	15.00	2.04	-
Género (M/F)	-	-	7/10	-	-	8/7
Anos de Escolaridade	9.24	2.14	-	8.87	2.36	-

Os sub-grupos apresentados na tabela anterior não apresentam diferenças estatisticamente significativas, no que respeita à idade (p=.353), ao género (χ^2=.473; p=.492) e aos anos de escolaridade (p=.646).

Variáveis em Estudo

Para a realização do estudo, como variáveis dependentes consideramos as provas neuropsicológicas, nomeadamente a Figura Complexa de Rey (cópia e memória) (FCRc e FCRm), a Procura da Chave (PC), o *Stroop* (as lâminas: Palavras, Cores e Interferência) (StroopP, StroopC e StroopI, respectivamente), o Código (parte B) (Cd), a Memória de Dígitos (directos e inversos) (DD e DI), o *Trail Making Test* (parte A e B) (TMT – A e TMT – B, respectivamente) e a Memória Lógica (ML).

As variáveis independentes referem-se ao diagnóstico de CC (TF e CIV) e às variáveis neonatais (Índice de Apgar 1 e 2, Perímetro Cefálico, Comprimento e Peso).

Instrumentos de Avaliação

A avaliação neuropsicológica consistiu na administração de provas, com o intuito de avaliar neurocognitivamente os sujeitos da amostra. A selecção das mesmas foi suportada por critérios que permitissem avaliar um maior número de funções cognitivas, no menor tempo possível. Desta forma, foi criado um protocolo de avaliação, no qual constavam as provas com as quais todos os participantes do estudo foram avaliados.

As provas foram as seguintes: Figura Complexa de Rey (cópia e memória); a prova Procura da Chave, da bateria *Behavioural Assessment of the Dysexecutive Syndrome* (BADS); o *Stroop* (as lâminas: Palavras, Cores e Interferência), as provas Código (parte B) e Memória de Dígitos (directo e inverso), da Escala de Inteligência de Wechsler para Crianças - III *(*WISC-III); o *Trail Making Test* (parte A e B) e a subprova Memória Lógica I da Escala de Memória de Wechsler - III (WMS-III).

A Figura Complexa de Rey é constituída por duas tarefas, a cópia e a reprodução por memória de um desenho geométrico complexo. A cópia do desenho permite avaliar a actividade perceptiva e a reprodução por memória permite avaliar a memória visual (Spreen & Strauss, 1998; Maia, Correia & Leite 2009). Como referido anteriormente, o instrumento consiste num desenho geométrico complexo, em que é solicitado ao sujeito que o copie e, após três minutos, é lhe novamente solicitado que o reproduza de memória. A pontuação é atribuída de acordo com a riqueza e exatidão da cópia e reprodução por memória, bem como os respectivos tempos de execução (Rey, 2002).

A Procura da Chave consiste num quadrado reproduzido numa folha de papel, que representa um campo, no qual é solicitado ao sujeito que desenhe o trajecto para procurar a chave que perdeu no referido campo. Esta prova permite avaliar concepção de estratégias de acção e resolução de problemas (Barbosa & Monteiro, 2008). A prova é avaliada e cotada de acordo com a funcionalidade e a probabilidade de sucesso, na procura da chave (Wilson, Alderman, Burgees, Emslie & Evans, 1996).

O Stroop – Teste de Cores e Palavras, é constituído por 3 lâminas, cada uma com 100 elementos, dispostos em cinco colunas de 20 elementos. A lâmina 1 consiste numa folha com as palavras "vermelho", "verde" e "azul" impressas na cor preta, o qual

é solicitado ao sujeito que as leia. A segunda lâmina consiste em conjuntos de "XXX" impressos a cor azul, vermelho ou verde, as quais o sujeito tem que nomear. A terceira lâmina é de Interferencia e é constituída pelas palavras azul, vermelho ou verde, impressas a cor não correspondente às mesmas. A pontuação é efectuada de acordo com o número de itens que o sujeito menciona em 45 segundos (Golden, 1994). Este teste permite avaliar a atenção selectiva (Spreen *et al.*, 1998).

A sub-prova Código tem como finalidade avaliar a velocidade de execução, o desempenho psicomotor e a organização perceptiva (Simões, 2002; Kamphaus, 2005).É constituída por uma folha de papel, onde estão impressos um conjunto de símbolos associados a números. É pedido ao sujeito que copie, por baixo de cada número, o símbolo correspondente. A duração da prova são 120 segundos, e a pontuação é dada consoante o número de símbolos correctamente associados ao número, neste tempo limite.

A sub-prova Memória de Dígitos é constituída por duas tarefas que podem ser administradas independentemente uma da outra. Os dígitos em ordem directa, que avaliam atenção auditivo-verbal, é constituído por várias sequências de números, lidas em voz alta pelo examinador. Seguidamente é pedido ao sujeito que repita a sequência pela mesma ordem que lhe foi apresentada. Os dígitos em ordem inversa, que avaliam a memória de trabalho, são igualmente constituídos por varias sequências de números, lidas pelo examinador. No entanto, é pedido ao sujeito que repita pela ordem inversa do que lhe foi apresentado.

Em ambas as tarefas, cada item é composto por dois ensaios. É atribuído um ponto, se o sujeito repetir correctamente uma das sequências do mesmo item, dois pontos se repetir correctamente duas sequências do mesmo item, e zero pontos se falhar em ambas as sequências. A tarefa é interrompida sempre que o sujeito falhe as duas sequências do mesmo item. A pontuação final corresponde á soma dos pontos obtidos em cada ensaio (Lezak, 1995; Simões, 2002).

A prova Trail Making Test é constituída por duas partes, A e B. A primeira é representada por uma folha de papel, onde estão dispostos aleatoriamente, um conjunto de círculos numerados de 1 a 25 (1-2-3-4-…), os quais o sujeito tem de unir, de forma

sequencial e por ordem crescente. Esta tarefa permite avaliar a capacidade de orientação visuo-espacial e a velocidade psicomotora (Lezak, 1995).

A segunda parte da prova (parte B) consiste num conjunto de números e letras, distribuídos numa folha de papel, os quais o sujeito deve unir por ordem crescente, de forma a alternar número e letra. Esta tarefa permite avaliar os processos atencionais, memória de trabalho e o funcionamento executivo, nomeadamente, planeamento, inibição e flexibilidade cognitiva (Demakis, 2004; Cangoz, Karakoc, Selekler, 2009).
A pontuação em ambas as partes é efectuada de acordo com o tempo despendido na sua realização. (Spreen & Strauss, 1998; Periáñez *et al*., 2007).

A Memória Lógica consiste numa prova, em que é efectuada a leitura de uma história, por parte do examinador, em que posteriormente é pedido ao sujeito que refira o maior número de conteúdos que se recorda. A pontuação é efectuada de acordo com a precisão em que o sujeito é capaz de repetir a história (unidade da história), sendo atribuído um ponto por cada unidade da história corretamente evocada e zero pontos por cada unidade omitida ou incorretamente evocada. Apresenta como finalidade avaliar a memória verbal imediata (Wechsler, 2008).

Procedimento Metodológico

Na realização do estudo, a administração dos testes neuropsicológicos ocorreu em dois contextos distintos.

A avaliação neuropsicológica do grupo de controlo, foi desenvolvida em contexto escolar, em escolas do distrito do grande Porto. Primeiramente foi requerida e aceite a autorização à direcção das escolas e posteriormente foi entregue um questionário aos pais, a fim de serem excluídos problemas neuropsiquiátricos e sistémicos. Assim, foi efectuada a recolha dos dados, que decorreu em contexto fechado.

Em contexto hospitalar foi realizada a avaliação neuropsicológica do grupo clínico, que decorreu no Hospital de São João – Porto. De igual forma, numa fase inicial foi solicitado e aceite o consentimento do Conselho de Ética do Hospital, garantindo

assim todas as directivas incluídas nas convenções internacionais para que os direitos dos pacientes fossem devidamente respeitados no decorrer da investigação.

Foi então iniciada a recolha dos dados do grupo clinico, que decorreu na consulta externa do serviço de Cardiologia Pediátrica, em contexto de gabinete fechado.

Os pacientes eram abordados na sala de espera, onde as investigadoras explicavam o objectivo da investigação, bem como a importância da sua participação no estudo. Após o parecer positivo, os participantes eram encaminhados para um gabinete onde eram realizadas as provas que constavam do protocolo de avaliação.

Em ambos os contextos de avaliação, foi entregue um consentimento informado aos pais e/ou adolescentes, onde era descrita a participação voluntária, as questões de confidencialidade e a possibilidade de desistência em qualquer momento.

Desenho

O desenho do estudo é transversal, pois a avaliação neuropsicologia foi realizada num único momento.

Para a recolha de informações retrospectivas do historial médico dos pacientes do grupo clínico, foi fundamental e necessária a colaboração da equipa administrativa do serviço em questão, que possibilitou o acesso ao arquivo dos processos clínicos dos mesmos.

Procedimento Estatístico

Com o intuito de efectuar a análise estatística dos dados anteriormente recolhidos, recorreu-se ao programa informático IBM SPSS *Statistic Editor* versão 20.0.

Numa primeira fase, no sentido de caracterizar a amostra, bem como descrever os resultados obtidos nas provas neuropsicológicas, procedeu-se a um estudo de carácter descritivo, com a análise das frequências e medidas de tendência central (média e desvio padrão). Para tal, foram consideradas variáveis demográficas (idade, género e anos de

escolaridade) e clínicas (Grupo de cardiopatias Congénitas e sub-grupos de Cardiopatias Congénitas, especificamente, TF e CIV), dos participantes em questão.

Posteriormente, foi realizada a comparação dos resultados obtidos pelos grupos (Grupo de Controlo e Grupo de Cardiopatias Congénitas) e sub-grupos de Cardiopatias Congénitas, nas provas neuropsicológicas. Para este efeito, utilizamos o teste não paramétrico U de *Mann-Whitney*.

Adicionalmente, procedeu-se a uma análise inferencial não paramétrica, onde foram efectuadas correlações de *Spearman* entre as variáveis neonatais e os resultados obtidos nas provas neuropsicológicas pelo grupo e pelos dois subgrupos de Cardiopatias Congénitas.

Para a análise estatística realizada, foram consideradas diferenças estatisticamente significativas de acordo com um nível de significância inferior ou igual a 0.05 ($p \leq 0.05$).

Capítulo V - Resultados

A análise dos resultados inerentes ao estudo será apresentada nas tabelas seguintes.

Primeiramente serão apresentados os resultados relativos ao desempenho dos dois grupos nas provas neuropsicológicas.

De acordo com a tabela 3, é possível verificar que o grupo de controlo apresenta melhores resultados em todas as provas, excepto na prova Cd.

Tabela 3

Resultados médios obtidos pelos dois grupos (GC e GCC) nas provas neuropsicológicas.

	GC		GCC	
	N=16		N=32	
	M	DP	M	DP
DD	12.56	1.41	8.84	2.49
DI	8,19	1,56	3,97	1,82
FCRc	35,63	0,62	30,03	8,57
FCRm	26,38	3,36	18,25	8,82
Cd	44,00	4,18	47,13	18,93
PC	11,19	2,69	6,66	3,29
StroopP	100,31	12,80	74,25	18,04
StroopC	76,94	12,01	61,34	13,65
StroopI	53,50	8,85	37,88	9,93
TMT – A	21,13	7,59	54,13	32,00
TMT - B	40,75	9,40	99,66	51,11
ML	13,13	2,34	10,81	4,84

No que respeita aos sub-grupos de Cardiopatias, podemos constatar que o sub-grupo CIV apresenta um melhor desempenho em todas as provas neuropsicológicas do que o sub-grupo TF (tabela 4).

Tabela 4

Resultados médios obtidos pelos dois sub-grupos de CC (sub-grupo CIV e sub-grupo TF).

	CIV N=17		TF N=15	
	M	DP	M	DP
DD	9,65	2,62	7,93	2,05
DI	4,35	1,84	3,53	1,77
FCRc	31,00	8,50	28,93	8,81
FCRm	20,56	8,06	15,63	9,18
Cd	50,12	21,47	43,73	15,59
PC	7,29	3,44	5,93	3,06
StroopP	78,12	19,18	69,87	16,17
StroopC	66,47	12,41	55,53	12,99
StroopI	40,71	11,06	34,67	7,60
TMT – A	48,24	33,62	60,80	29,76
TMT - B	87,24	51,79	113,73	48,16
ML	11,29	5,27	10,27	4,43

A *tabela 5* estabelece uma comparação dos resultados obtidos nas provas neuropsicológicas pelos dois grupos. Desta forma, verificamos que existem diferenças estatisticamente significativas entre os dois grupos, no que respeita ao desempenho em todas as provas neuropsicológicas ($p \leq .001$), excepto na prova Cd e ML.

Tabela 5

Comparação dos resultados obtidos pelos dois grupos (GC e GCC), nas provas neuropsicológicas.

	GC	GCC		
	N=16	N=32		
	Mean Rank	Mean Rank	U	*p*
DD	**37,88**	**17,81**	**42,000**	**< .001**
DI	**39,31**	**17,09**	**19,000**	**< .001**
FCRc	**35,66**	**18,92**	**77,500**	**< .001**
FCRm	**33,59**	**19,95**	**110,500**	**.001**
Cd	23,22	25,14	276,500	.654
PC	**36,16**	**18,67**	**69,500**	**< .001**
StroopP	**36,56**	**18,47**	**63,000**	**< .001**
StroopC	**33,97**	**19,77**	**104,500**	**.001**
StroopI	**37,03**	**18,23**	**55,500**	**< .001**
TMT – A	**10,50**	**31,50**	**480,000**	**< .001**
TMT - B	**10,91**	**31,30**	**473,500**	**< .001**
ML	29,25	22,12	180,000	.095

Na *tabela 6* é apresentada a comparação dos resultados obtidos nas provas neuropsicológicas pelos dois sub-grupos de Cardiopatias Congénitas. Assim, verificamos que existem diferenças estatisticamente significativas entre os dois sub-grupos, no que concerne ao desempenho neuropsicológico nas provas DD (*p*=.049), StroopC (*p*=.018), StroopI (*p*=.024) e TMT – B (*p*=.049).

Tabela 6

Comparação dos resultados obtidos pelos dois sub-grupos de CC (Grupo CIV e Grupo TF), nas provas neuropsicológicas.

	CIV N=17 Mean Rank	TF N=15 Mean Rank	U	*p*
DD	**19,56**	**13,03**	**75,500**	**.049**
DI	18,41	14,33	95,000	.230
FCRc	18,06	14,73	101,000	.331
FCRm	18,79	13,90	88,500	.142
Cd	17,41	15,47	112,000	.576
PC	18,06	14,73	101,000	.331
StroopP	18,85	13,83	87,500	.132
StroopC	**20,15**	**12,37**	**65,500**	**.018**
StroopI	**19,97**	**12,57**	**68,500**	**.024**
TMT - A	13,79	19,57	173,500	.082
TMT - B	**13,44**	**19,97**	**179,500**	**.049**
ML	17,59	15,27	109,000	.502

Na *tabela 7* são apresentados os valores médios relativamente às variáveis neonatais dos dois sub-grupos de Cardiopatias. Podemos verificar que o sub-grupo CIV revela valores médios superiores nas variáveis Índice de Apgar 2, Perímetro Cefálico e Peso e um valor médio inferior na variável Índice Apgar 1, do que o sub-grupo TF. Relativamente à variável comprimento, ambos os sub-grupos apresentam um valor médio semelhante.

Tabela 7

Valores médios relativos às variáveis neonatais dos dois sub-grupos de CC.

	CIV N=14		TF N=11	
	M	DP	M	DP
Índice de Apgar (1)	7,75	2,70	7,77	2,20
Índice de Apgar (2)	9,88	0,34	9,46	0,97
Perímetro Cefálico	34,81	2,07	32,45	3,03
Comprimento	45,92	11,56	45,92	5,98
Peso	3,31	0,50	2,79	0,79

Com o intuito de verificar a existência de uma correlação significativa entre as variáveis neonatais e o desempenho cognitivo nas provas neuropsicológicas administradas, no grupo e subgrupos de cardiopatias congénitas, utilizou-se a correlação ordinal de Spearman (rho), uma vez que, a distribuição da amostra não é homogénea.

Através da análise da *tabela 8* verifica-se que, no grupo das cardiopatias congénitas, as variáveis neonatais correlacionam-se de forma positiva e estatisticamente significativa com as seguintes provas neuropsicológicas:

- O Indice de Apgar 1 com o StroopI (ρ=.370; p=.048);

- O Indice de Apgar 2 com os DD (ρ=.389; *p*=*.037*), a FCRc (ρ=.370; *p*=.048*)*, o StroopP (ρ=.482; *p*=.008), o StroopC (ρ=.476; *p*=.009) e com o StroopI (ρ=.516; *p*=.004);
- O Perímetro Cefálico com os DD (ρ=.453; *p*=.023), a PC (ρ=.446; *p*=.025), o StroopC (ρ=.603; *p*=.001) e com o StroopI (ρ=.672; *p*<.001);
- O Comprimento com o StroopP (ρ=.547; *p*=.023);
- O Peso com os DD (ρ=.559; *p*=.001), a FCRc (ρ=.403; *p*=.027), a FCRm (ρ=.434; *p*=.016), a PC (ρ=.473; *p*=.008), o StroopP (ρ=.393; *p*=.032), o StroopC (ρ=.542; *p*=.002) e o StroopI (ρ=.571; *p*=.001);

Ainda neste grupo constatamos que a variável Peso se correlaciona de forma negativa e estatisticamente significativa com a prova TMT – B (ρ=-.403; *p*=.027).

Tabela 8

Correlação de Spearman entre as variáveis neonatais (Índice de Apgar 1 e 2, Perímetro Cefálico, Comprimento e Peso) e os resultados obtidos nas provas neuropsicológicas no Grupo CC.

	Índice Apgar (1)		Índice Apgar (2)		Perímetro Cefálico		Comprimento		Peso	
	ρ	*p*	ρ	*p*	ρ	*p*	ρ	*p*	ρ	*p*
DD	.046	.814	**.389**	**.037**	**.453**	**.023**	.264	.167	**.559**	**.001**
DI	.059	.762	.171	.374	.367	.071	.021	.913	.325	.080
FCRc	-.080	.679	**.370**	**.048**	.350	.087	.350	.062	**.404**	**.027**
FCRm	-.269	.158	.187	.330	.285	.167	.137	.480	**.434**	**.016**
Cd	.008	.965	.108	.578	.261	.208	.290	.127	.252	.180
PC	-.021	.915	-.094	.627	**.446**	**.025**	.095	.623	**.473**	**.008**
StroopP	.223	.244	**.482**	**.008**	.389	.054	**.547**	**.002**	**.393**	**.032**
StroopC	.172	.371	**.476**	**.009**	**.603**	**.001**	.304	.109	**.542**	**.002**

StroopI	**.370**	**.048**	**.516**	**.004**	**.672**	**<.001**	.335	.076	**.571**	**.001**
TMT - A	-.137	.478	-.203	.291	-.341	.096	.011	.954	-.343	.064
TMT - B	-.167	.386	-.244	.201	-.386	.057	-.094	.629	**-.403**	**.027**
ML	-.182	.345	.040	.838	.151	.470	.100	.605	.277	.138

De acordo com a *tabela 9* é possível verificar que, no subgrupo CIV, as variáveis neonatais correlacionam-se de forma positiva e estatisticamente significativa com as diferentes provas neuropsicológicas, nomeadamente:

- O Perímetro Cefálico com a PC (ρ=.651; *p*=.012), o StroopC (ρ=.720; *p*=*.004*) e com o StroopI (ρ=.740; *p*=.002);
- O Comprimento com o StroopP (ρ=.552; *p*=.027).
- O Peso com os DD (ρ=.507; *p*=.045), a PC (ρ=.700; *p*=.003) e com o StroopI (ρ=.526; *p*=.036).

Tabela 9

Correlação de Spearman entre as variáveis neonatais e os resultados obtidos nas provas neuropsicológicas no Subgrupo CIV.

	Índice Apgar (1)		Índice Apgar (2)		Perímetro Cefálico		Comprimento		Peso	
	ρ	*p*	ρ	*p*	ρ	*p*	ρ	*p*	ρ	*p*
DD	-.031	.909	.187	.487	.364	.201	.350	.184	**.507**	**.045**
DI	.045	.869	-.021	.939	.316	.272	-.156	.564	.050	.853
FCRc	-.004	.989	.335	.204	.279	.334	.373	.154	.306	.249
FCRm	-.314	.236	.123	.650	.371	.192	.151	.577	.424	.101
Cd	-.317	.232	.000	1.000	.237	.414	.342	.195	.279	.295
PC	-.080	.769	.083	.760	**.651**	**.012**	-.047	.864	**.700**	**.003**
StroopP	.040	.882	.411	.114	.333	.244	**.552**	**.027**	.429	.097
StroopC	-.089	.744	.144	.595	**.720**	**.004**	.190	.480	.475	.063
StroopI	.306	.249	.474	.064	**.740**	**.002**	.266	.319	**.526**	**.036**
TMT - A	-.230	.392	-.205	.446	-.476	.085	-.209	.438	-.430	.096
TMT – B	-.084	.756	-.164	.544	-.348	.223	.438	-.119	-.483	.058
ML	-.150	.578	.021	.940	.130	.659	.003	.991	.340	.198

Pela análise da *tabela 10* verificar-se que, no subgrupo TF, as variáveis neonatais correlacionam-se de forma positiva e estatisticamente significativa com as provas neuropsicológicas, nomeadamente:

- O Indice de Apgar 2 com o StroopC (ρ=.601; *p*=.030);
- O Peso com os DD (ρ=.561; *p*=.037).

Tabela 10

Correlação de Spearman entre as variáveis neonatais e os resultados obtidos nas provas neuropsicológicas no Subgrupo TF.

	Índice Apgar (1)		Índice Apgar (2)		Perímetro Cefálico		Comprimento		Peso	
	ρ	*p*	ρ	*p*	ρ	*p*	ρ	*p*	ρ	*p*
DD	.226	.459	.470	.105	.437	.179	.194	.526	**.561**	**.037**
DI	.106	.731	.205	.501	.416	.203	.225	.460	.491	.075
FCRc	-.145	.636	.234	.442	.256	.447	.326	.276	.474	.087
FCRm	-.332	.268	-.061	.844	-.044	.898	-.014	.964	.240	.409
Cd	.305	.310	.114	.710	.194	.568	.193	.528	.222	.446
PC	.015	.962	-.314	.296	.216	.524	.181	.555	.120	.684
StroopP	.432	.140	.508	.076	.284	.397	.446	.127	.299	.299
StroopC	.419	.154	**.601**	**.030**	.312	.351	.372	.210	.458	.100
StroopI	.494	.086	.464	.111	.513	.107	.374	.208	.509	.063
TMT - A	-.077	.802	-.003	.991	.148	.664	.373	.209	-.132	.652
TMT - B	-.140	.649	-.084	.785	-.212	.531	.052	.865	-.257	.375
ML	-.247	.415	-.095	.758	.086	.802	.227	.455	.190	.515

Capítulo VI – Discussão dos Resultados

Após análise cuidada dos resultados obtidos, torna-se fundamental reflectir sobre eles, de modo a verificar a importância e magnitude dos mesmos em relação à literatura até então existente.

De acordo com os resultados obtidos, quando comparamos a performance nas provas neuropsicológicas do grupo de adolescentes com CC com o grupo de adolescentes saudáveis, verificamos que o primeiro apresenta um desempenho inferior em quase todas as dimensões neuropsicológicas estudadas, excepto em Cd e ML, sendo assim confirmada a hipótese I. De acordo com a literatura existente, de facto as crianças com cardiopatias congénitas, demonstram um baixo desempenho na avaliação neuropsicológica, em vários domínios cognitivos (Miatton *et al*, 2007a).

O desempenho do grupo de CC demostrou a existência de dificuldades ao nível da atenção auditivo-verbal imediata (prova DD), atenção selectiva (prova StroopP, StroopC e StroopI) e atenção dividida (prova TMT - B). Este facto é corroborado por estudos anteriores, em que os pacientes com cardiopatias congénitas obtiveram desempenhos inferiores em tarefas que requerem domínios atencionais (Miatton *et al*, 2007a; Daliento *et al*, 2005; Brosig *et al*, 2007; Gerdes & Flynn, 2010) e pelos resultados obtidos na avaliação comportamental efectuada pelos pais de crianças com cardiopatias congénitas, onde descrevem problemas atencionais por parte dos filhos (Shillingfor et al, 2008).

No que respeita à prova PC, verificamos que os adolescentes com cardiopatias congénitas apresentam um pior desempenho. Este dado sugere dificuldades ao nível do funcionamento executivo, nomeadamente na capacidade de planeamento. Os resultados são corroborados por estudos que referem a existência de alterações no funcionamento executivo, principalmente na capacidade de planeamento, organização e resolução de problemas (Bellinger *et al*, 2010; Miatton *et al*, 2007a; Daliento *et al*, 2005; Majnemer *et al*, 2008). As alterações no funcionamento executivo também são consistentes com os baixos resultados na prova TMT – B.

De acordo com os resultados obtidos, verificamos que os adolescentes com cardiopatias congénitas apresentam défices ao nível da capacidade visuo-construtiva

(prova FCRc) e visuo-espacial (prova FCRMc e TMT - A). Segundo Miatton *et al*, 2007a; Brosig *et al*, 2007; Bellinger *et al*, 2003b; Bellinger *et al*, 2010, crianças com diferentes tipos de CC, revelam dificuldades em tarefas que envolvam a capacidade visuo-espacial. Os défices visuo espaciais, parecem estar implicados em alterações ao nível da organização perceptiva (Bellinger *et al*, 2003a).

Os nossos resultados, demostraram também que o grupo de cardiopatias congénitas apresenta um pior desempenho nas provas que avaliam a memória visual (prova FCRm) e a memória de trabalho (prova DI). Existe uma consistência destes resultados com alguns estudos existentes na literatura, onde mencionam que crianças com cardiopatias congénitas apresentam défices de memória (Bellinger *et al*, 2003b; Miatton *et al.,* 2008; Majnemer *et al*., 2008), sobretudo ao nível da memória de trabalho (Bellinger *et al.*, 2003b) e memória visual (Tindall, Rothermel, Delamater, Pinsky, Klein, 1999). Tindall e colaboradores (1999) referem que crianças com cardiopatias não apresentam alterações na memória verbal, sendo este dado igualmente consistente com o presente estudo. No entanto, outros estudos mencionam que crianças com cardiopatias não apresentam défices de memória, quando comparadas com crianças saudáveis (Hövels-Gürich, Seghaye, Sigler, Kotlarek, Bartl *et al.*, 2001).

Na presente investigação foram também efectuadas comparações relativas ao desempenho nas provas neuropsicológicass entre os sub-grupos de cardiopatias congénitas, nomeadamente cardiopatia cianótica (grupo TF) e cardiopatia acianótica (grupo CIV).

Relativamente às provas DD, StroopC e StroopI, que avaliam a atenção auditivo-verbal imediata e a atenção selectiva, respectivamente, apuramos que o grupo de TF apresenta piores resultados, comparativamente ao grupo de CIV. Segundo Hovels-Gurich e colaboradores (2007), as crianças com cardiopatias congénitas cianóticas apresentam um maior risco de disfunção atencional comparativamente com as crianças com cardiopatias acianóticas. Estes resultados sugerem então que a hipótese II foi parcialmente confirmada.

No que concerne ao funcionamento executivo, avaliado pela prova TMT – B, apuramos que o grupo TF, apresenta piores resultados do que o grupo CIV.

Contudo, apesar das diferenças encontradas no desempenho neuropsicológico entre os subgrupos acianótico e cianótico, estes dados não são totalmente consistentes

com a literatura, onde não se verificam diferenças na performance neuropsicológica entre os grupos (Miatton et al, 2007b; Oates, Simpson, Cartmill & Turnbull, 1995).

A presente investigação apresentou também como objectivo correlacionar variáveis neonatais e o desempenho cognitivo nas provas neuropsicológicas no grupo e sub-grupos de cardiopatias congénitas.

Relativamente ao Grupo CC verificou-se que existem correlações positivas em todas as variáveis neonatais com diferentes provas neuropsicológicas. Especificamente, verificou-se que o Índice de Apgar 1 apresenta uma correlação positiva com a atenção selectiva e o Índice de Apgar 2 apresentou correlações positivas com a atenção auditivo-verbal imediata, atenção selectiva e capacidades visuo-construtivas.

Recém-nascidos com Cardiopatias Congénitas tendem a apresentar lesões hipoxico-isquémicas (Massaro, Glass, Brown, Chang, Krishnan, Jonas & Donofrio, 2011), detetadas através de ressonância magnética, em recém nascidos com cardiopatias congénitas (Block, McQuillen, Chau, Glass & Poskitt, 2010), podendo estas estarem relacionadas com baixos valores no Índice de Apgar, repercutindo-se mais tarde no desempenho cognitivo destas crianças (Massaro, Glass, Brown, Chang, Krishnan, Jonas & Donofrio, 2011).

Na variável Perímetro Cefálico, existem correlações positivas com a atenção auditivo-verbal imediata, a capacidade de planeamento e a atenção selectiva.

O perímetro cefálico parece ser indicador de um baixo desempenho em várias tarefas cognitivas implicadas no lobo frontal. Através de mecanismos de auto-regulação da circulação sanguínea, algumas áreas cerebrais são mais protegidas do que outras. As artérias cerebrais anteriores apresentam uma melhor resposta auto-reguladora, sendo que a redistribuição do fluxo sanguíneo favorece a perfusão dos lobos frontais. No entanto, em situações de comprometimento da oxigenação fetal, pode tornar-se prejudicial, como no caso da presença de CC (Donofrio & Massaro, 2010).

Na variável comprimento, existe uma correlação positiva com a atenção selectiva, avaliada pela prova StroopP. No que respeita à variável Peso, existe uma correlação positiva com a atenção auditivo-verbal, capacidade visuo-construtiva, memoria visuo-construtiva, capacidade de planeamento e atenção selectiva. Ainda na

variável peso, verificamos uma correlação negativa com a atenção alternada, capacidade de planeamento e flexibilidade cognitiva.

Como mencionado anteriormente, em casos de comprometimento de oxigenação, ocorre uma restrição global do crescimento somático, devido à redistribuição da circulação cerebral (McQuilen & Miller, 2010). Assim, um atraso no crescimento fetal, poderá influenciar o desempenho neurocognitivo a longo prazo.

Estes resultados podem ser corroborados por estudos anteriores que relacionam o baixo peso à nascença com o comprometimento das capacidades cognitivas (Böhm, Katz-Salamon, Smedler, Lagercrantz & Forssberg, 2002; Gaynor, Wernovsky, Jarvik, Bernbaum, Gerdes, Zackai, Nord, Clancy, Nicolson & Spray, 2007).

Pela análise destes resultados podemos verificar que todas as variáveis neonatais se correlacionam de forma positiva com a atenção selectiva, podendo ser considerado que existe uma forte influência das variáveis neonatais ao nível dos processos atencionais. Segundo Gaynor e colaboradores (2006), as variáveis neonatais como o Perímetro Cefálico, Peso, Índice de Apgar (1º e 5º minutos) podem ser indicadores da vulnerabilidade no índice de desenvolvimento mental e psicomotor em crianças.

De acordo com os resultados acima descritos, foi possível verificar que a variável peso, exerce uma forte influência no desempenho neuropsicológico, sendo que se correlacionou com grande parte das provas em questão. Segundo (Böhm, *et al*., 2002), os défices cognitivos estão relacionados com o baixo peso à nascença.

No que concerne ao sub-grupo CIV, verificamos que existem correlações positivas entre o perímetro cefálico e a capacidade de planeamento e com a atenção selectiva. Alguns autores sugerem que crianças com Cardiopatias Congénitas apresentam baixo perímetro cefálico, com consequências a longo prazo, ao nível neurodesenvolvimental (Wernovsky, 2006; Donofrio & Massaro, 2010), sendo que o nosso estudo, remete para as consequências ao nível do domínio atencional.

Ainda neste grupo, existem correlações positivas entre o comprimento e a atenção selectiva e o Peso e a atenção auditivo-verbal, a capacidade de planeamento e a atenção selectiva. Estes resultados são suportados pelo padrão de restrição do crescimento somático em caso de comprometimento de oxigenação fetal.

No sub-grupo TF, verificam-se correlações positivas entre o Índice de Apgar 2 e a atenção selectiva e o Peso e a atenção auditivo-verbal.

Recém-nascidos com Cardiopatias Congénitas tendem a apresentar lesões hipoxico-isquémicas (Massaro, Glass, Brown, Chang, Krishnan, Jonas & Donofrio, 2011), detetadas através de ressonância magnética, em recém nascidos com cardiopatias congénitas (Block, *et al*., 2010), podendo estas estarem relacionadas com baixos valores no Índice de Apgar, repercutindo-se mais tarde no desempenho cognitivo destas crianças (Massaro, *et al*., 2011).

No entanto, pudemos constatar que o grupo TF, foi aquele onde existiram menos correlações entre as variáveis neonatais e o desempenho neurocognitivo. Segundo Miatton e colaboradores (2007b), o baixo peso em crianças com TF, exerce influência no desempenho neuropsicológico, sendo demonstrada no presente estudo, ao nível do dominio atencional.

Assim, de acordo com estes resultados é possível confirmar as hipóteses previamente colocadas (Hipóteses III, IV e V), visto existirem correlações entre as variáveis neonatais e as diferentes provas neuropsicológicas no grupo e sub-grupos de cardiopatias congénitas.

Capítulo VII – Conclusão

O objectivo da investigação era verificar o impacto da presença de Cardiopatia Congénita no desempenho cognitivo dos adolescentes, bem como verificar se variáveis neonatais estavam relacionadas com o desenvolvimento neuropsicológico dos mesmos.

À condição crónica da doença cardíaca congénita, surgem associadas diversas limitações que impõem uma multiplicidade de problemas relacionados com a qualidade de vida, o ajustamento psicossocial e o desenvolvimento cognitivo desta população.

Apesar dos avanços no diagnóstico e tratamento da patologia, que aumentaram a esperança de vida destes pacientes, torna-se de facto imperativo actuar ao nível dos problemas supra mencionados, de modo a minimizar o impacto da doença e garantir o desenvolvimento ideal nestas crianças, já que estes défices poderão persistir na idade adulta.

Como pudemos verificar, esta população encontra-se ainda em idade escolar. Desde logo, os resultados encontrados permitiram perceber, que estes adolescentes apresentam diversas dificuldades ao nível neurocognitivo, que por sua vez assumem um impacto agravado ao nível do sucesso escolar. Neste sentido torna-se urgente perceber quais os factores que poderão estar na origem deste tipo de problemática.

Desde o momento da concepção até ao momento do nascimento, ocorrem inúmeras e sobretudo determinantes alterações, capazes de desenvolver um ser tão frágil e indefeso. No entanto, na presença de patologia o desenvolvimento é comprometido a vários níveis. Assim, foi importante verificar no nosso estudo, a relação entre as variáveis neonatais e o funcionamento cognitivo. De facto, verificamos que as variáveis ao nascimento influenciam a longo prazo, o desenvolvimento neurocognitivo em vários domínios. O comprometimento dos mesmos poderá estar relacionado com o anormal crescimento intrauterino, que na presença de CC repercute-se no desenvolvimento anormal das várias áreas cerebrais e do crescimento somático.

Por tudo isto, é importante a continuidade do estudo destas relações, de modo a robustecer as nossas conclusões e aperfeiçoar a compreensão de quais os domínios cognitivos que apresentam maior vulnerabilidade às variáveis neonatais.

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Anexos

Anexo 1

Consentimento Informado

TERMO DE CONSENTIMENTO INFORMADO

Eu, _____, depois de ter sido integralmente informado dos objectivos e âmbito do Projecto de Investigação intitulado "Implicações neurocognitivas das Cardiopatias Congénitas", declaro que aceito participar neste estudo.

Além de ter sido garantida a confidencialidade dos dados recolhidos, fui também informado de que, em caso de não aceitar participar neste projecto, não ocorrerão quaisquer consequências na minha assistência médica habitual.

Porto, ____ de _____ de 201_

Assinatura: _____

Anexo 2

Instrumentos

B

Exemplo

2	1	4	6	3	5	2	1	3	4	2	1	3	1	2	3	1	4	2	6	3

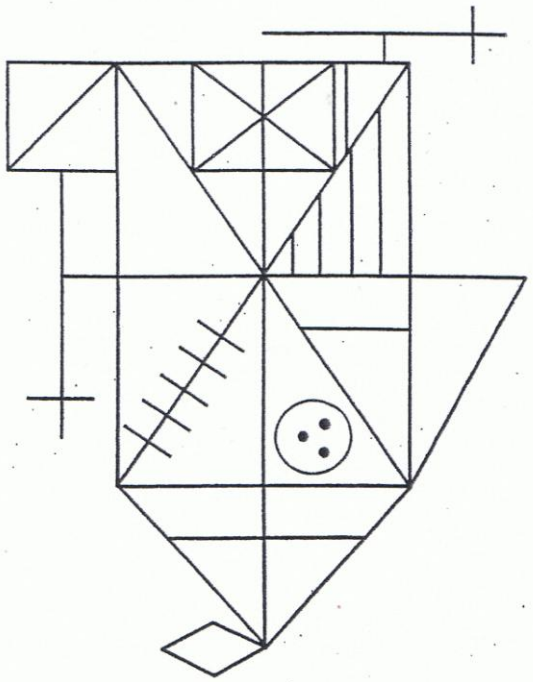
1	2	5	1	3	1	5	4	2	7	4	6	9	2	5	8	4	7	6	1	8

7	5	4	8	6	9	4	3	1	8	2	9	7	6	2	5	8	7	3	6	4

5	9	4	1	6	8	9	3	7	5	1	4	9	1	5	8	7	6	9	7	8

2	4	8	3	5	6	7	1	9	4	3	6	2	7	9	3	5	6	7	4	5

2	7	8	1	3	9	2	6	8	4	1	3	2	6	4	9	3	8	5	1	8



11. Pesquisa de Símbolos

Interromper após 120 segundos

	6-7 Parte A	8-16 Parte B
Tempo limite	120"	120"
Tempo Despendido		
Nº de itens correctos		
Nº de itens incorrectos		
Pontuação Total Obtida (máximo = 45)		

12. Memória de Dígitos

Administrar ambos os ensaios de cada item, inclusive quando o Ensaio 1 é bem sucedido
Interromper após 2 insucessos consecutivos
Administrar os Dígitos em Sentido Inverso inclusive quando o sujeito obteve uma pontuação de 0 nos Dígitos em Sentido Directo

Dígitos em Sentido Directo					
Idade do sujeito	Ensaio 1 / Resposta	Cotação	Ensaio 2 / Resposta	Cotação	Pontuação final 0, 1
1	2 - 9		4 - 6		
2	3 - 8 - 6		6 - 1 - 2		
3	3 - 4 - 1 - 7		6 - 1 - 5 - 8		
4	8 - 4 - 2 - 3 - 9		5 - 2 - 1 - 8 - 6		
5	3 - 8 - 9 - 1 - 7 - 4		7 - 9 - 6 - 4 - 8 - 3		
6	5 - 1 - 7 - 4 - 2 - 3 - 8		9 - 8 - 5 - 2 - 1 - 6 - 3		
7	1 - 6 - 4 - 5 - 9 - 7 - 6 - 3		2 - 9 - 7 - 6 - 3 - 1 - 5 - 4		
8	5 - 3 - 8 - 7 - 1 - 2 - 4 - 6 - 9		4 - 2 - 6 - 9 - 1 - 7 - 8 - 3 - 5		
				Total Dígitos em Sentido Directo (máximo = 16)	

Dígitos em Sentido Inverso					
Idade do sujeito	Ensaio 1 / Resposta	Cotação	Ensaio 2 / Resposta	Cotação	Pontuação final 0, 1
	Exemplo: 8 - 2		Exemplo: 5 - 6		
1	2 - 5		6 - 3		
2	5 - 7 - 4		2 - 5 - 9		
3	7 - 2 - 9 - 6		8 - 4 - 9 - 3		
4	4 - 1 - 3 - 5 - 7		9 7 - 8 - 5 - 2		
5	1 - 6 - 5 - 2 - 9 - 8		3 6 - 7 - 1 - 9 - 4		
6	8 - 5 - 9 - 2 - 3 - 4 - 2		4 - 5 - 7 - 9 - 2 - 8 - 1		
7	6 - 9 - 1 - 6 3 - 2 - 5 - 8		3 - 1 - 7 9 - 5 - 4 - 8 - 2		
				Total Dígitos em Sentido Inverso (máximo = 14)	

Pontuação Total Obtida (máximo = 30)

13. Labirintos

Interromper após 2 insucessos consecutivos, excluindo o Labirinto 1. Aos sujeitos com idades entre 8 anos administrar os Labirintos 1 a 3 em caso de insucesso ou sucesso parcial no Labirinto 4. Caso o su obtenha 0 pontos no Labirinto 4, fazer a demonstração do Labirinto Exemplo e administrar os Labir 1 a 3.

	Labirintos	Tempo Limite	Tempo Despendido	Número de erros	Cotação Rodear com um círculo a pontuação obtida.	Pontu
6-7	Exemplo					
	1	30"			2+ erros 0 / 1 erro 1 / 0 erros 2	
	2	30"			2+ erros 0 / 1 erro 1 / 0 erros 2	
	3	30"			2+ erros 0 / 1 erro 1 / 0 erros 2	
8-16	4	30"			2+ erros 0 / 1 erro 1 / 0 erros 2	
	5	45"			2+ erros 0 / 1 erro 1 / 0 erros 2	
	6	60"			2+ erros 0 / 1 erro 1 / 0 erros 2	
	7	120"			3+ erros 0 / 2 erros 1 / 1 erro 2 / 0 erros 3	
	8	120"			4+ erros 0 / 3 erros 1 / 2 erros 2 / 1 erro 3 / 0 erros 4	
	9	150"			4+ erros 0 / 3 erros 1 / 2 erros 2 / 1 erro 3 / 0 erros 4	
	10	150"			5+ erros 0 / 4 erros 1 / 3 erros 2 / 2 erros 3 / 1 erro 4 / 0 erros 5	
					Pontuação Total Obtida	

Recomendações para o subteste Labirintos (ver Capítulo 6 do Manual) (máximo = 28)

As seguintes recomendações podem ser fornecidas, caso sejam necessárias, mas cada uma delas só poderá ser feita uma vez no decurso da administração do subteste

1 *"Deves começar aqui."* (apontar para o centro do quadrado)
2 *"Não podes passar através de uma parede."*
3 *"Não pares. Continua até encontrares a saída. Podes voltar para trás."*
4 *"Não começes outra vez. Continua a partir daqui* (apontar para o ponto atingido) *e tenta encontrar o caminho certo para sores."*

Nome _____ *Data* _____

WISC-III™

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2 Memória Lógica I

Registo:
Assinalar (✓) cada Unidade de História literalmente evocada. Em cada Unidade de História, registar as respostas não literais.

Cotação:
0 ou 1 ponto por cada unidade, de História ou Temática. Consultar os critérios de cotação descritos no Manual de Administração e Cotação (Capítulo 4 e Anexo A).

História A

Maria Lopes, que vive no Lumiar em Lisboa e que trabalha como cozinheira no refeitório de uma escola, queixou-se na esquadra da polícia de ter sido assaltada na Avenida da Liberdade, na noite anterior, e de lhe terem roubado cinquenta e seis euros. Tinha quatro filhos pequenos, a renda por pagar e não comiam há dois dias. A polícia, comovida com a história desta mulher, organizou um peditório em seu favor.

História A	Cotação: Unidade de História		Cotação: Unidade Temática		Critério de Cotação
Maria	0	1			É necessário referir *Maria*.
Lopes	0	1			É necessário referir *Lopes*.
que vive no Lumiar	0	1			*Lumiar* (em qualquer contexto).
em Lisboa	0	1			*Lisboa* (em qualquer contexto).
			0	1	Indicação de que a protagonista é do sexo feminino.
e que trabalha	0	1			Indicação de que tem um trabalho.
como cozinheira	0	1			É necessário referir *cozinheira*, ou outra forma da palavra.
no refeitório	0	1			É necessário referir *refeitório* ou sinónimo.
de uma escola,	0	1			É necessário referir *escola* ou sinónimo.
			0	1	Indicação de que a protagonista está empregada ou a trabalhar.
queixou-se	0	1			Indicação de que foi feita uma queixa formal à autoridade (em qualquer contexto).
na esquadra	0	1			*Esquadra* (em qualquer contexto) ou uma palavra ou frase que indique que é uma esquadra da polícia.
da polícia	0	1			*Polícia* (em qualquer contexto).
de ter sido assaltada	0	1			Indicação de que foi assaltada.
na Avenida da Liberdade,	0	1			Na *Avenida da Liberdade* (em qualquer contexto).
na noite anterior,	0	1			Indicação de que o assalto ocorreu na noite anterior.
e de lhe terem roubado	0	1			Indicação de que foi roubada.
cinquenta e seis euros.	0	1			Indicação de um valor em dinheiro superior a 49 e inferior a 60 euros.
			0	1	Indicação de que a protagonista queixou-se de ter sido roubada.
Tinha quatro	0	1			É necessário referir o número *quatro*, juntamente com a indicação de que as crianças eram suas.
filhos pequenos,	0	1			É necessário referir *filhos pequenos* ou sinónimos.
			0	1	Indicação de que a protagonista tem filhos.
a renda por pagar	0	1			Uma frase indicando que tem a renda de casa por pagar.
e não comiam	0	1			Indicação de que as crianças ou família estavam sem comer (sem comida).
há dois dias.	0	1			É necessário referir *dois dias*, ou uma frase que indique cerca de dois dias.
			0	1	Indicação de que as personagens encontram-se necessitadas e precisam de ajuda.
A polícia,	0	1			Uma palavra ou frase indicando um ou mais elementos da polícia (em qualquer contexto).
comovida com a história desta mulher,	0	1			Indicação de que a sua história provocou empatia nos outros.
			0	1	Indicação de que a polícia se comoveu com a história da mulher.
organizou um peditório	0	1			Uma frase indicando que houve recolha de dinheiro.
em seu favor.	0	1			Indicação de que o dinheiro era para a mulher e/ou para os seus filhos.
			0	1	Indicação de que a polícia respondeu de imediato/directamente às necessidades da protagonista.

História A
Pont. Evocação Unid. História
Mínimo = 0 Máximo = 25

História A
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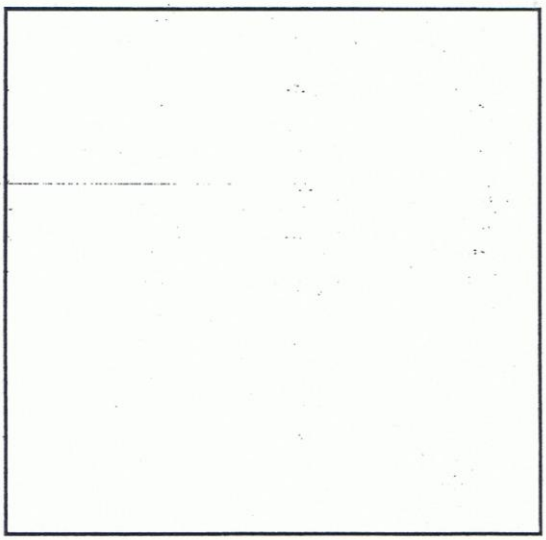
Mínimo = 0 Máximo = 7

Key Search Test

Subject's name



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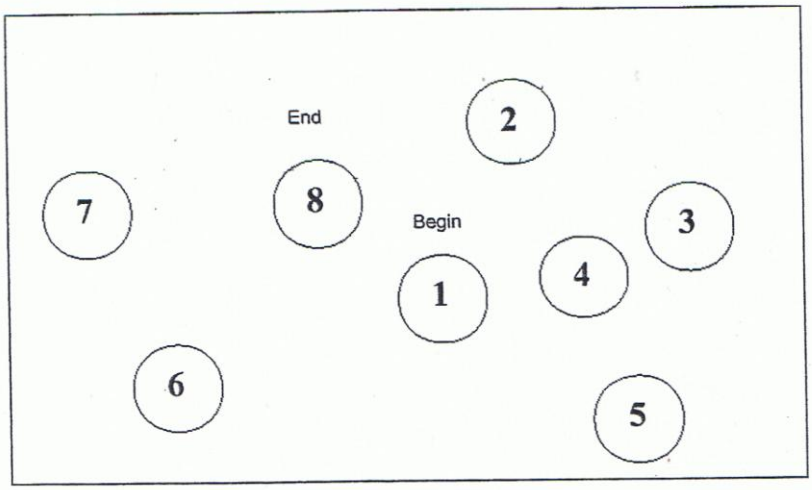
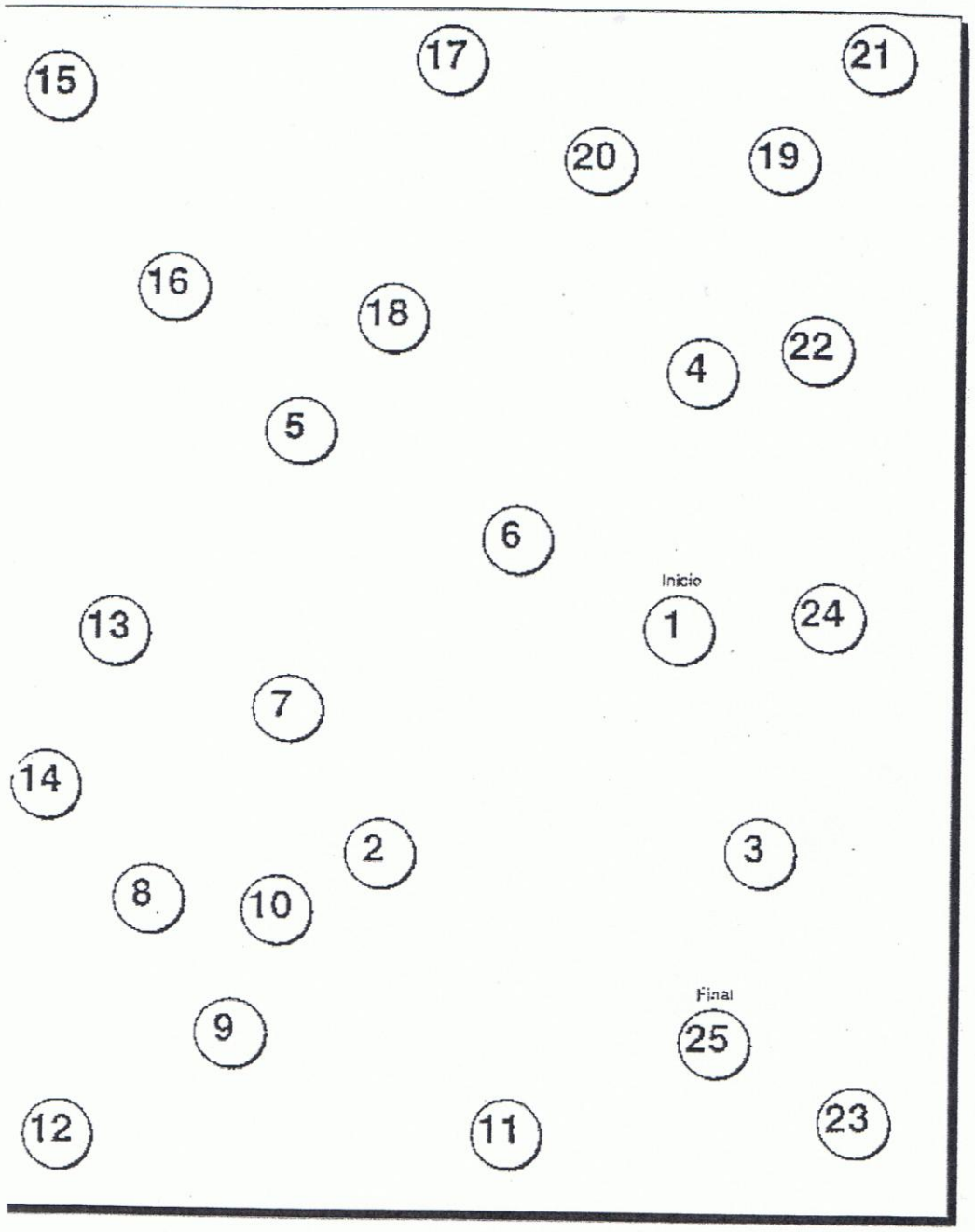
Trail Making Test Part A – *SAMPLE*

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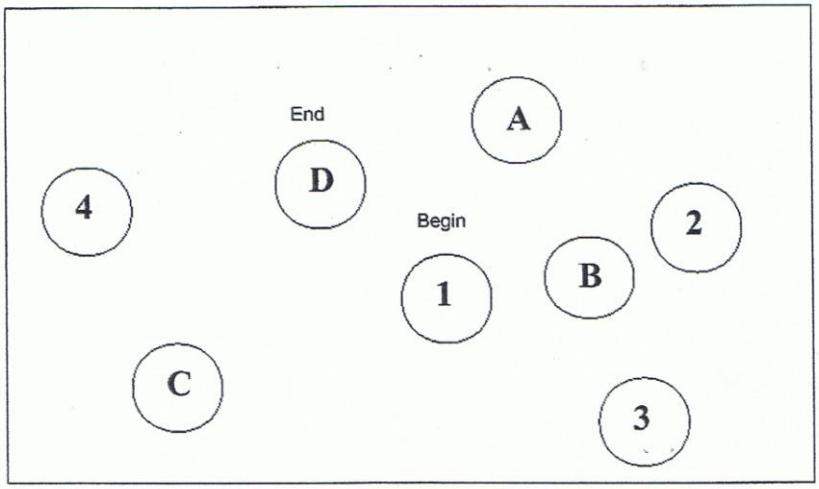
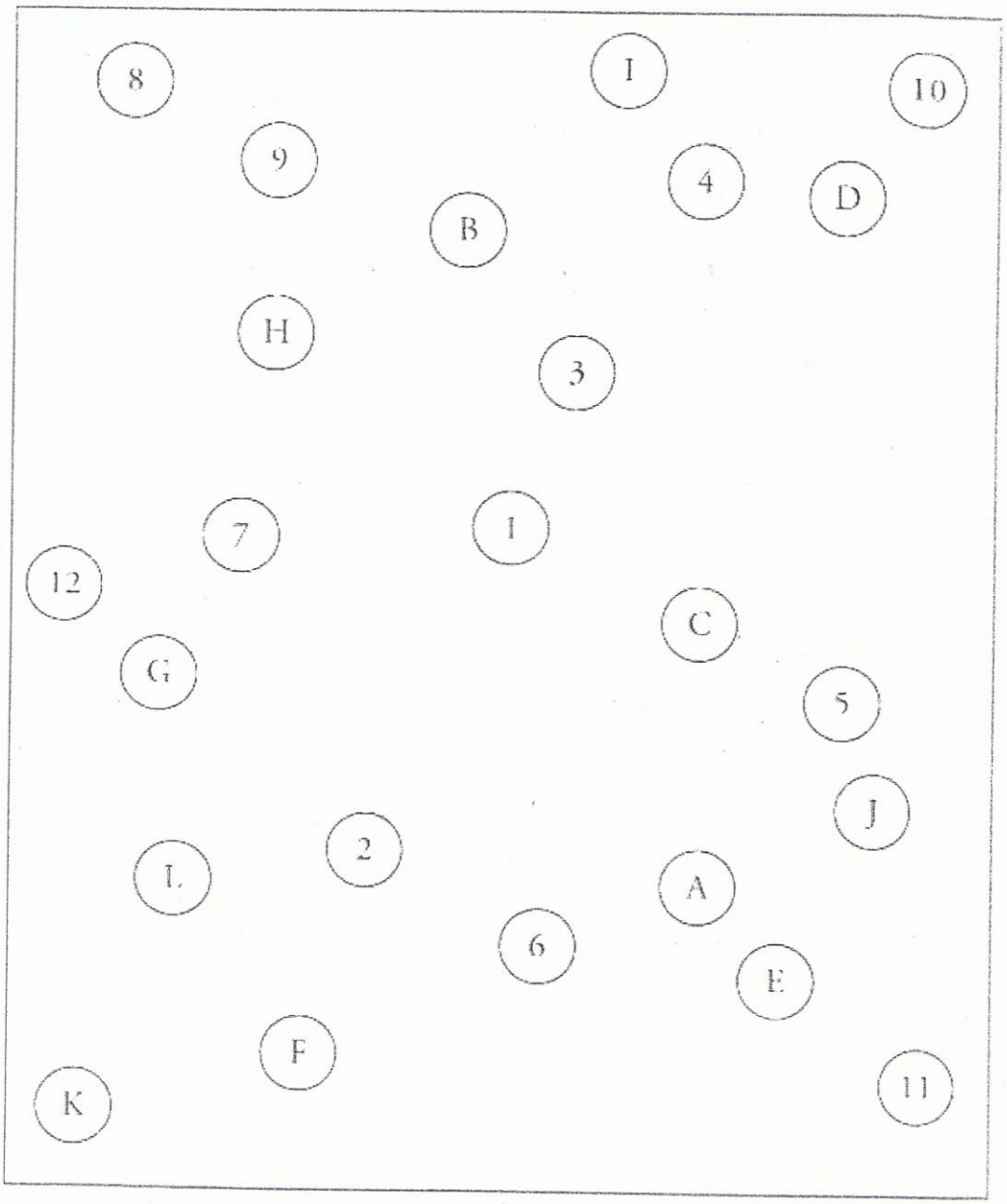
Trail Making Test Part B – *SAMPLE*

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Anexos

Abstracts submetidos a Congressos Internacionais

Anexo 1

Primeira autora de um resumo submetido e aceite em formato poster no congresso da American Academy of Pediatrics em New Orleans – 20/23 de Outubro 2012

How does fetal development affects neuropsychological abilities in adolescents with Congenital Heart Disease?

S. Sarmento 1, 2, S. Moreira 1, 2, S. Matos 1, 2, B. Peixoto 1, 2, J. C. Areias 3, 4, M. E. G. Areias 1, 5

1 Department of Psychology, ISCS-N (CESPU), 2.UNIPSA/ CICS (CESPU), GANDRA PRD, 3 Department of Pediatric Cardiology, Hospital S. João, 4 Oporto Medical School, University of Porto, Porto, 5 CINEICC, University of Coimbra, Coimbra, Portugal

Purpose: Patients with congenital heart disease (CHD) may show neuropsychological deficits in pre-school and school ages. Surgeries and anoxia have been studied as main variables in these disorders but recently the focus has been turned to fetal cardiac and circulatory conditions that may affect neurodevelopment. The purpose of this investigation was to evaluate the neurocognitive performance in adolescents with CHD and to determine whether parameters of fetal development evaluated in neonates, such as head circumference, length, weight and Apgar scores, are somehow related to their neurocognitive performance.

Methods: 61 CHD patients (34 males) aged from 13 to 18 years old (mean=15.08 ± 1.72), 37 cyanotic (20 Tetralogy of Fallot, 17 Transposition of the Great Arteries) and 24 acyanotic (Ventricular Septal Defect), enrolled in this study. We assessed also a control group with 16 healthy children (11 males and 5 females) ages ranging from 13 and 18 (mean=15.69 ± 1.44). All assessment measures for CHD patients were once obtained in a tertiary hospital; the control group was evaluated in school. Demographic information and clinical history were collected. Neuropsychological assessment included Wechsler's Digit Test (direct and reverse) and Symbol Search, Rey's Complex Figure, BADS's Key Searching Test, Color-Word Stroop Test, Trail Making Test and Logical Memory Task.

Results: CHD patients compared to control group showed lower scores in the Wechsler's Digit Test, in direct (u=192.000; p<0,001) and reverse (u=104.000; p<0.001) versions, in Rey's Complex Figure, copy (u=132.500; p<0,001) and memory (u=127.500; p<0.001), in Symbol Search (u= 650.000; p=0.042), in BADS's Key Searching Test (u= 128.000; p<0.001) in Stroop Test, words (u=119.000; p<0.001), colors (u=182.500; p<0.001) and interference (u=104,500; p<0.001) and in Trail Making Test, A (u=906.000; p<0.001) and B (u=895.500; p<0.001). **C**yanotic compared to acyanotic patients showed lower scores in Wechler's Digit Test, in direct form (u=593.500; p=0,026), in Rey's Complex Figure, memory (u=627.500; p=0.007), in Stroop Test, words (u=606.500; p=0.016), colors (u=633.000; p=0.005) and interference (u=645,000; p=0.003), and in Trail Making Test, A (u=285.000; p=0.019) and B (u=296.500; p=0.029). Several correlations were apparent between fetal/ neonatal parameters and neuropsychological abilities in each type of CHD. However, the circumference of the head at birth stands as a main correlation with cognitive development later on in all kinds of CHD (Direct Digit: rho=0.437, p=0.004; BADS: rho=0.346, p=0.021; Stroop Words: rho=0.366, p=0.014; Stroop Colours: rho=0.408, p=0.006; Stroop Interference: rho=0.397, p=0.008; Trail Making Test A: rho=0.390, p=0.009).

Conclusion: Adolescents with CHD have worse neuropsychological performance than the control group, and the cyanotic worse than the acyanotic patients. The fetal

anomalies in heart and circulation seem to have impact in delaying cerebral and somatic growth, predicting cognitive impairment in adolescents with CHD.

Anexo 2

Co - Autora de um Resumo Aprovado para Comunicação Oral no "World Congress of Cardiology" Scientific Sessions Abril de 2012 no Dubai

Psychosocial Adjustment of Patients with Congenital Heart Disease

I. Freitas 1, 2,*, M. Castro 1, 2, S. Sarmento 1, 2, F. Teixeira 1, 2, R. Coelho 1, 2, V. Viana 3, 4, J. C. Areias 5, 6, M. E. G. Areias 1, 7
1Department of Psychology, ISCS-N (CESPU), 2UNIPSA/ CICS (CESPU), GANDRA PRD, 3Hospital S. João, 4Faculty of Nutrition, University of Porto, 5Department of Pediatric Cardiology, Hospital S. João, 6Oporto Medical School, University of Porto, Porto, 7CINEICC, University of Coimbra, Coimbra, Portugal

Introduction: Progress in diagnosing, pharmacological treatment and surgery, has resulted in significantly improved survival rates among patients with Congenital Heart Disease (CHD).

Objectives: We aimed to study Psychosocial Adjustment (PSA), Psychiatric Morbidity (PM), Quality of Life (QOL), School Performance (SP), Physical Limitations (PL), and Social Support (SS) of adolescents and young adults with CHD.

Methods: We evaluated 99 CHD patients, 57 males, aged from 12 to 26 years old (M=18.12 ± 3.65), 55 cyanotic. The participants were interviewed on such topics as SS, family/educational background, self-image, PL and emotional adjustment, were administered a standardized psychiatric interview (SADS-L) and completed self-report questionnaires on QOL (WHOQOL-BREF) and PSA (YSR and ASR). Observational versions of the same questionnaires (CBCL, ABCL) were filled by one of their relatives. Full clinical and demographic history was collected.

Results: We found a 21% rate of lifetime prevalence of psychopathology (14% in males and 31% in females) and 49% of school retentions (M=1.52 years + 0.50). Patients with severe forms of CHD showed worse PSA than those with moderate and mild forms of illness (u=762.000; p=0.026), those submitted to surgery showed worse QOL on physical (t=- 2,396; p=0,019), psychological (t=-2,327; p=0,022), SR (t=-2,171; p=0,033) and general (u=534,500; p=0,040) dimensions, and worse PSA (more withdrawn: u=508,500; p=0,028). Participants without pharmacological therapy revealed better QOL in general domain (u= 1048,500; p=0,024). SS has a great impact improving patients' physical (t=2,752; p=0,007), psychological (t=3,396; p=0,001), SR (t=4,699; p=0,000), environment (t=2,805; p=0,006) and general (u=482,000; p=0,000) QOL and poorer SS resulted in more withdrawn (u=499,000; p=0,003) and social problems (u=577,500; p=0,022). Patients with more PL showed worse physical (t=-2,088; p=0,039), psychological (t=-2,607; p=0,011), SR (t=-2,267; p=0,026) and general (u=700,500; p=0,000) QOL and more withdrawn (u=783,500; p=0,006). Female patients showed more somatic complaints (u=205,000;p=0,037), anxiety/ depression (u=756,000;p=0,002), aggressive behavior (u=688,000;p=0,000), thought problems (u=824,000;p=0,007), internalization (u=716,000;p=0,001) and externalization (u=905,500; p=0,039). Good performance in school also showed a significant impact incrementing QOL and PSA.

Conclusion: The PSA and QOL of patients with CHD are crucial. SS, PL, and SP have a significant impact over them.

Anexo 3

Co-Autora de um Resumo Aprovado para Comunicação Oral no American Heart Association Scientific Sessions 2011 em Orlando – Florida
Circulation. 2011; 124: A15388

Living with a Congenital Heart Disease: Quality of Life, Psychosocial Adjustment, Psychiatric Morbidity and School Performance May be Affected, but Social Support Plays an Important Role in promoting Resilience

Maria Emilia G Areias[1]; Flávio Teixeira[2]; Rosália Coelho[2]; Isabela Freitas[2]; Marta Castro[2]; Sofia Sarmento[2]; Cidália Proença[2]; Ana Margarida Silva[2]; Daniela Vieira[2]; Cláudia Vaz[2]; Cláudia Moura[3]; Victor Viana[4]; José Carlos Areias[3]

[1] Psychology, Instituto Superior de Ciências da Saúde - Norte (CESPU); CINEICC, Gandra-Paredes, Portugal
[2] Psychology, Instituto Superior de Ciências da Saúde - Norte (CESPU); UNIPSA, Gandra-Paredes, Portugal
[3] Pediatrics (Cardiology), Porto Med Sch, Univ of Porto, Porto, Portugal
[4] Nutrition, Faculty of Nutrition, Univ of Porto, Porto, Portugal

Introduction: Good care of congenital heart disease (CHD) allows patients to survive until a later adulthood, facing challenges for adjustment throughout life.

Hypothesis: Quality of Life (QOL) and Psychosocial Adjustment (PSA) are affected in some CHD patients. They are more likely to have psychopathological disorders and more prone to school failures. Social support (SS) is a protecting variable, promoting resilience. The aims of this investigation were to study QOL, Psychiatric Morbidity (PM), PSA, School Peformance (SP), Physical Limitations (PL), and SS of adolescents and young adults with CHD.

Methods: 86 CHD patients, 48 male, aged from 12 to 26 years (M= 18,52 ± 3,70), 51 cyanotic. Clinical and demographic history was collected. Participants were interviewed once on topics as SS, family educational style, self-image, PL and emotional adjustment, administered a psychiatric interview (SADS-L) and completed self-report questionnaires on QOL (WHOQOL-BREF) and PSA (YSR and ASR). One of their relatives filled the observational versions of the questionnaires (CBCL, ABCL).

Results: We found a 22,1% lifetime prevalence rate of psychopathology and 51.2% retentions in school (M= 1,68 year + 0.829). Comparing to normal population, our patients have better QOL in environmental (t=4.327; p=0.000) and social relationships (t=2.795; p=0.006) dimensions. Cyanotic patients showed worse QOL in environmental dimension (t=-2.120; p=0.037); Complex CHD reported more social (u = 600.000; p = 0.028), thought (u = 607.000; p =0.031) and externalization (u=586.500; p=0.021) problems. Patients who had surgery reported worse QOL in social relationships dimension (t=-2.506; p=0.014) and overall (t=-2.107; p=0.038), and more withdrawn behavior (u=303.500; p=0.009). Those with better SS revealed better QOL in psychological (t=2.893; p=0.005), social relationships (t=2.521; p=0.014), environment (t=2.871; p= 0.005) and physical (t=2.734; p=0.008) dimensions, and less withdrawn behavior (u=963.000;p=0.004) and social problems (u=904.500; p=0.024).

Conclusions: CHD patients seemed to be more prone to PM, worse PSA and SP, although SS plays a crucial role in all variables and in resilience.

Key Words: Quality of life • Behavioral aspects • Adult congenital heart disease • Congenital heart disease

Anexo 4

Co-Autora de um Resumo Convidado para Comunicação Oral no "Fetal and Pediatric Cardiology Seminar" em Paris –10 Dezembro 2011

Living with Congenital Heart Disease: Quality of Life of Patients with Congenital Heart Diseases

Areias MEG (1, 3), Teixeira F (1,4), Coelho R (1,4), Freitas, I (1,4) Castro, M (1,4), Sarmento, S (1,4), Proença C (1,4), Silva AM (1,4), Vieira D (1,4), Vaz C (1,4), Moura C (2,5,6) ; Viana V (2, 4), Areias JC (2, 5)

(1) Department of Psychology of ISCS-N (CESPU), Gandra, Portugal

(2) Department of Pediatrics (Cardiology), Porto Medical School, UP, Porto, Portugal

(3) CINEICC, Coimbra, Portugal

(4) UNIPSA, Gandra, Portugal

(5) Unidade de Investigação Cardiovascular, Porto, Portugal

(6) Hospital São João

Anexo 5

Co-Autora de uma Apresentação por Convite no 46th Annual Meeting of Association for European Paediatric and Congenital Cardiology em Istambul - Maio 2012

Living with Congenital Heart Disease: Quality of Life in Early Adult Life

Areias MEG (1,3), Freitas I (1,4), Castro M(1,4), Sarmento S(1,4), Pinto C (1,4), Vieira P (1,4), Matos S(1,4), Viana V (6,7), Areias JC (2,5,6)

(1) Department of Psychology of ISCS-N (CESPU), Gandra, Portugal

(2) Department of Pediatrics (Cardiology), Porto Medical School, UP, Porto, Portugal

(3) CINEICC, Coimbra, Portugal

(4) UNIPSA, Gandra, Portugal

(5) Unidade de Investigação Cardiovascular, Porto, Portugal

(6) Hospital São João

(7) Faculty of Nutrition, University of Porto

Anexo 6

Co-Autora de um Resumo Aprovado para Comunicação Oral no "Quality of Care & Outcomes Research in CV Disease & Stroke", Scientific Sessions 2012 em Atlanta – Georgia

Circ Cardiovasc Qual Outcomes. 2012; 5: A93

How is the Quality of Life and the Psychosocial Adjustment affected in Patients with Congenital Heart Disease?

Castro, M (1,4), Freitas, I (1,4), Sarmento, S (1,4) Teixeira F (1, 4), Coelho R (1, 4), Viana V (2, 4), Areias JC (2, 5), Areias MEG (1, 3)

(1) Department of Psychology of ISCS-N (CESPU), Gandra, Portugal

(2) Department of Pediatrics (Cardiology), Porto Medical School, UP, Porto, Portugal

(3) CINEICC, Coimbra, Portugal

(4) UNIPSA, Gandra, Portugal

(5) Unidade de Investigação Cardiovascular, Porto, Portugal

Introduction: High rates of survival in Congenital Heart Disease (CHD) allowed patients to face different challenges in life cycle, and made the topics on adjustment and quality of life more and more central in healthcare.

Hypothesis: We tested the hypothesis that CHD has a negative impact over psychosocial adjustment, psychiatric morbidity, quality of life and school performance, and that the severity of disease and the number of surgeries increase the negative impact over adjustment and the social support generates a buffer, good effect on it.

Objective: We aimed to study Quality of Life (QOL), Psychosocial Adjustment (PSA), Psychiatric Morbidity, School Performance, Physical Limitations, and Social Support of adolescents and young adults with CHD.

Methods: We evaluated 110 CHD patients, 62 males, aged from 12 to 26 years old (M=18.00 ± 3.62), 58 cyanotic. The participants were interviewed on such topics as social support, family/educational background, self-image, physical limitations and emotional adjustment, were administered a standardized psychiatric interview (SADS-L) and completed self-report questionnaires on QOL (WHOQOL-BREF) and PSA (YSR and ASR). Observational versions of the same questionnaires (CBCL, ABCL) were filled by one of their relatives. Full clinical and demographic history was collected.

Results: We found a 22% rate of lifetime prevalence of psychopathology (14.5% in males and 31.3% in females) and 50% of school retentions (M=1.50 years + 0.50). Patients with severe forms of CHD showed worse PSA than those with moderate and mild forms of illness (internalization: u=939.000; p=0.030), the cyanotic versus acyanotic and those with moderate-to-severe residual lesions versus mild ones have worse QOL on physical dimension; those submitted to surgery showed worse QOL on physical (t=-2.525; p=0.013), psychological (t=-2.394; p=0,018), social relationships (t=-2.502; p=0,014) and general (u=1294,000; p=0.006) dimensions, and worse PSA (more withdrawn: u=719,000; p=0,037). Social support has a great impact improving patients' physical (t=2.707; p=0,008), psychological (t= 2.755; p=0.007), social relationships (t=4.976; p=0,000), environment (t=3.085; p=0,003) and general (u=623.500; p=0,000) QOL and poorer social support resulted in more withdrawn (u=557.500; p=0.000) and social problems (u=748.500; p=0,023). Patients with more physical limitations showed worse physical (t=-2.093; p=0,039), psychological (t=-2.674; p=0.009) and general (u=971.500; p=0,002) QOL and more withdrawn (u=1023.000; p=0,015). Female patients showed more somatic complaints (u=260.000;p=0,011), anxiety/ depression (u=984.000;p=0,002), aggressive behavior (u=920.500;p=0,001), thought problems (u=1069.500;p=0,010), internalization (u=869.000;p=0,000) and externalization (u=1163.000; p=0,050). Good performance in school also showed a significant impact incrementing QOL and PSA.

Conclusion: We concluded that we should set a special emphasis in maximizing social support and attention in improving school performance, when supplying care in CHD, as they have a positive impact over self-confidence of patients and adjustment.

Anexo 7

Comunicação a convite da Faculdade de Psicologia e Ciências da Educação da Universidade de Coimbra CINEICC "Conversas com Investigação" 29 de Março de 2012

Viver com uma Cardiopatia Congénita: Ajustamento psicossocial, morbilidade psiquiátrica e qualidade de vida em adolescentes e jovens adultos com cardiopatias congénitas; Aspetos do desenvolvimento em doentes com patologia cianótica e acianótica.

Areias MEG (1,3), Freitas I (1,4),Castro M (1,4), Sarmento S (1,4), Moura C (2,5,6) Viana V (6,7), Peixoto B (1,4) Areias JC (2,5,6)

(1) Department of Psychology of ISCS-N (CESPU), Gandra, Portugal

(2) Department of Pediatrics (Cardiology), Porto Medical School, UP, Porto, Portugal

(3) CINEICC, Coimbra, Portugal

(4) UNIPSA, Gandra, Portugal

(5) Unidade de Investigação Cardiovascular, Porto, Portugal

(6) Hospital São João

(7) Faculty of Nutrition, University of Porto

Anexos

Artigos submetidos a Revistas com Peer Review

Anexo 1

Primeira Autora de Um artigo pronto a submeter ao "American Heart Journal"

How does fetal development affects neurodevelopment in young adults with Congenital Heart disease?

Maria Emília Guimarães Areias,
Rua Central de Gandra, 1317,
4585-116 Gandra PRD – Portugal
T:+351 224 157 176
F:+351 224 157 102
metega@sapo.pt ; memilia.areias@cespu.pt

Sofia Sarmento [1,4]

Sara, Moreira [1,4]

Isabela Freitas [1,4]

Marta Castro [1,4]

Bruno Peixoto[1,4]

Victor Viana [2,4]

José Carlos Areias [2,5]

Maria Emília Guimarães Areias [1,3]

[1]Department of Psychology, Instituto Superior de Ciências da Saúde do Norte (CESPU), Gandra, Portugal
[2]Department of Pediatrics (Cardiology), Hospital São João, Porto Medical School, University of Porto, Portugal
[3]CINEICC, Centro de Investigação do Núcleo de Estudos e Intervenção Cognitivo-Comportamental, Coimbra, Portugal
[4]UNIPSA, Unidade de Investigação de Psicologia e Saúde/ CICS (CESPU), Gandra, Portugal
[5]Unidade de Investigação Cardiovascular da Faculdade de Medicina do Porto, Portugal

ABSTRACT

Background: It is believed that in-uterus alterations may have an effect on later neurodevelopment of children with congenital heart disease. The purpose of this research is to evaluate the neurocognitive performance in adolescents with congenital heart disease (CHD) and to understand if the neonatal variables such as Apgar scores; head circumference, the length and weight, are somehow related with the neurocognitive performance of these adolescents.

Methods: We evaluated 70 CHD patients (40 males) aged from 13 to 18 years old (mean=15.14 ± 1,71), 39 being cyanotic. As for the control group, we had 16 healthy children (11 males) with ages between 13 and 18 (mean=15.69 ± 1.44). All assessment measures were once obtained in a tertiary hospital and the control group were evaluated in their schools. Demographic information and clinical history were collected. For neuropsychological assessment, different tasks were used, such as Wechler's Digit Test, Wechler's Symbol Search, Rey's Complex Figure, Key Search Test, Color-Word Stroop Test, Trail Making Test and Logical Memory Task.

Results: CHD patients, when compared to the control group, showed lower scores in the Wechler's Digit Test, in direct (u=219.000; p<0,001) and in the indirect (u=«134.500; p<0.001); Rey's Complex Figure, copy (u=149.000; p<0,001) and memory (u=134.000; p<0.001); Wechler's Symbol Search (u=778.000; p=0,016); Key Search Test (u= 153.500; p<0.001); Stroop Test, words (u=131.000; p<0.001), colors (u=211.500; p<0.001) and interference (u=123.000; p<0.001); Trail Making Test, A (u=1.033.000; p<0.001) and B (u=1.000.000; p<0.001).

Conclusions: Adolescents with CHD have worse neuropsychological performance than the control group, and the cyanotic worse than the acyanotic patients. The fetal anomalies in heart and circulation seem to have impact in delaying cerebral and somatic growth, predicting cognitive impairment in adolescents with CHD.

INTRODUCTION

Congenital heart disease has been considered as the most common cause of child morbidity and mortality.[1]

Despite all advances in surgical interventions, this may be a contributor to the increase of life expectancy of these children, over half of these will develop some kind of neurological damage.[2]

Many children that were submitted to cardiac surgery in the neonatal period, show a pattern of neurological problems throughout their development, such as, cognitive deficits, attention problems and executive functioning, visual-motor e visual-spacial skills , delay in expressive language acquisition and learning difficulties. [3,4,5,6,7,8]

Until today, studies about neurological development in children with congenital heart disease, have only been focused on factors related to surgeries, such as cerebral perfusion being compromised during extra corporal circulation.[1] Although, other studies have found a high percentage of neurological abnormalities, anatomical and functional, before surgery in newborn babies with congenital heart disease[9,10,11] and also found a connection between these abnormalities and cognitive development throughout childhood.

According to some studies, neurobehavioral abnormalities, such as hypotonic, agitation, motor asymmetry, lethargy and autistic characteristics, were found prior to surgery in over 50% of newborns (less than a month old) and 38% of children (between one month old and 2 years old) with congenital heart disease. These abnormalities usually persist or get worse in the postoperative period, were cranial nerve damage may also occur.[12,13]

Still during the postoperative period, other complications may also occur, being the most common ones characterized as agenesis or disgenesis of corpus callosum, holoprosemcephaly, microcephaly, lissencephaly,[14] incomplete closure of the operculum, ischemic changes and white substance lesion caracterized by periventricular leukomalacia (PVL). [15]

Newborns with congenital heart disease show higher risk of developing hypoxia, hypotension and acidosis. This may due to the fact that congenital cardiac malformations and abnormalities in fetal growth are related.[16,11] Other hypothesis suggest that abnormalities in cerebral development, inter-uterus hemodynamic alterations and congenital cerebral abnormalities and acquired cerebral lesions, linked

to prolonged cyanosis and hipoperfusion after birth,[1] also contribute to the neurological development difficulties in children with congenital heart disease.[9]

Newborns with congenital heart disease show high levels of acquired cerebral lesions, proved by studies using postoperative magnetic resonance imaging scans. The high number of cases of newborns with lesions in the white substance suggests that there is a vulnerability that might be related with the delay in brain development. These abnormalities in newborns with congenital heart disease may reflect trouble in cerebral blood flow in the fetus.[17]

For many newborns, congenital heart disease seems to be an isolated anomaly, believing that the brain has potential for a normal development.[18] Although brain and heart development occur simultaneously in the fetus with congenital heart disease, being the first morphogenetic programs of each organs share the same genetic vias.

In the seventh week of the gestation period the heart is considered to be morphologically mature, but brain development will continue on, as different morphological events occur throughout the first two trimesters,[19] such as neuronal migration and arborization, sinaptogenesis, programmed cellular death, oligodrendocits maturation and reorganization or synaptic connections.[9]

Following this period there is a constant brain growth and is directly linked with the formation and perfectioning of connections in the third trimmest and neonatal period. [19] This development is associated with the increase in metabolic activity were the brain relies on the heart for oxygen and nutrient supply.

The existence of congenital heart disease will increase the chances of blood flow abnormalities in the fetus, resulting in the compromised brain development due to the complex relations between common cells, genetic programming, physiological consequences of cerebral blood flow alterations and the dynamic of oxygen distribution during brain development.[17] This can be seen as good evidence that these factors take an important role in cerebral growth.[20]

In what is considered as normal fetal circulation, gas exchange occur through the placenta. The deoxygenized blood coming from the vena cava goes directly in the right ventricle and through the arterial canal to the placenta. The Eustachian valve and the arterial septum move together to get the venous blood from the hepatic inferior vena cava to the right ventricle and the oxygenated blood from the venous canal over the oval foramen through the left ventricle to the aorta and cerebral circulation.

When the fetal oxygenation is compromised there is a blood redistribution to the cerebral circulation, this phenomenon is known as brain sparing,[1] resulting in a

pattern of global distribution of the somatic growth, with preservation of the head growth.[17]

This hemodynamic growth is represented by the diastolic flow in the cerebral arteries and the decrease of diastolic flow in the descending and umbilical aorta.[1] It is believed that many brain areas may be better protected than others. According to Dubiel, Gunnarsson & Gunnarsson[21], a study with pregnant women with complications in their pregnancy due to maternal hypertension and placenta dysfunction, cerebral vasodilatation was found in 41% of women in the anterior cerebral artery, 30% in the posterior cerebral artery and 24% in the medium cerebral artery. In this way, anterior cerebral arteries show a better auto regulative response, were the redistribution of blood flow is favorable to the perfusion of the frontal lobes. Although, medium cerebral arteries are presented as less reactive.[1]

This mechanism has been found to be a contributor to an adverse neurological development, since the cerebral vasodilatation occurs when there is a compromise in the fetal oxygenation. This protection mechanism doesn't seem to be enough to keep a normal brain development and growth in prolonged stress situations in the uterus.[1]

In a normal fetal blood circulation, the oxygenated blood is taken to the brain and the deoxygenized blood goes to the placenta.[1]

As said previously, studies show abnormalities in blood flow, that occur in congenital heart disease for instance, such as Hipoplasic Left Heart Sindrome, Transposition of the Great Arteries, Tetralogy of Fallot, may contrinute to an abnormal brain development.[9,1,17]

In fetus with Hipoplasic Left Heart Syndrome there may be an increase in the resistance to cerebral flow, where the blood flow returns through an istmo-aortic hipoplasia in order to get to the brain.

In fetus with Transposition of the Great Arteries, the venous blood from the cerebral circulation goes back directly to the brain.

When Tetralogy of Fallot and Hipoplasic Right Heart Syndrome, is found in fetus, the deoxygenized blood goes in the cerebral circulation due to the intracardiac mix.

This allows the statement that the type of lesion does not only affects the cerebral blood flow but also the degree of deoxygenized blood distributed in the cerebral circulation. [1]

All alterations in the blood flow may be related to some sort of abnormal somatic growth in uterus, having an influence in the neonatal anthropometry. [16]

When comparing to newborns without congenital heart disease, newborns with Hipoplasic Left Heart Sindrome, show lower weight, length and head circumference smaller than normal and the head volume is disproportionally low when compared to their weight. Newborns with Transposition of the Great Arteries show normal weight, but a lower head circumference when compared to their weight. On the other hand, newborns with Tetralogy of Fallot have normal proportions, although their weight, length and head circumference are smaller than expected. Children with Coarctation of the Aorta have higher head volume, when compared to their weight at birth.[1,17,22,16]

Many factors are believed to be related to the low weight at birth, including genetic syndromes, placenta insufficiency, and inter-uterine growth restriction. All these may increase the risk of neurodevelopment delays.

Recently, Gaynor, Jarvik, Bernbaum, Gerdes, Wernovsky et al [23] showed that, in general, factors inherent to the patient, such as weight at birth, head circumference at birth and the Apgar score in the first and fifth minute, better explain the vulnerability of mental and psychomotor growth than the intraoperative factors (weight in surgery, cooling time, deep hypothermic circulatory arrest time, cardiopulmonary bypass time, lowest nasopharyngeal temperature).

Methods

Participants

The study enrolled 70 CHD patients, 40 male and 30 female, with a mean age of 15.14 ± 1.71 years (range:13-18 years old); while the control group had 11 males and 5 females with mean age of 15.69 ± 1.44.

When it comes to ages, these two groups have no differences between them (F=1.387; p=0.242); as for academic performance (F=3.474; p=0.066); and also between gender (χ^2=0.727; p=0.394).

O GCC was still divided in 3 sub groups, the sub group TF (n=21; 13 male and 8 female),the subgroup CIV(n=29; 14 male and 15 female), and TGA (n=20; 13 male and 7 female).

Between these three groups there are no differences when it comes to age (F=0.178; p=0.837); academic performance (F=0.236; p=0.791) and gender (χ^2=1.630; p=0.443).

As for educational background, patients with CHD showed a mean of 9.16 (± 1.97) and the control group showed a mean of 10.13 (± 1.36). For 39 individuals the CHD

was cyanotic and for 31 acyanotic; when analyzed by their age, cyanotic patients showed a mean of 15.08 (± 1.87) while acyanotic had a mean of 15.23 (± 2.15). As for their gender, we had 14 female cyanotic patients and 25 males; while 16 females and 15 males had acyanotic type of CHD.

Instruments

In order to collect all necessary information for this study, all participants underwent a brief neuropsychological assessment, designed to evaluate a number of neurocognitive functions over a short period of time.

Different tests were used for this purpose such as, Wechler's Digit Test, in direct and indirect form, focused on the assessment of immediate auditory-verbal attention and working memory, respectively; Wechler's Symbol Search, used to evaluate psychomotor performance, speed of execution, perceptive organization and persistence. Rey's Complex figure, copy and reproduction from memory three minutes after image exposure, was used in order to assess visual constructional ability and visual constructional memory. The Key Search Test, from the Behavioral Assessment of the Dysexecutive Syndrome- children, focuses on the evaluation of the capability for efficient planning.

Color-word Stroop Test was used to assess attention efficiency. Trail Making Test, part A focuses on the evaluation of visual spatial orientation, psychomotor speed, while part B is ment to assess divided attention.

Finally, Wechler's Logical Memory Task was used in order to evaluate verbal memory.

Procedure

During this research, patients were approached to participate in this study while waiting for their cardiology or pediatric cardiology appointment. At this time, they were informed about all aspects of the research being asked to sign a consent, signed by the patients themselves or their caregivers when under 18 years old.

The data analysis of the different instruments was processed using the software IBM SPSS (Statistical Package for the Social Sciences) Statistics, version 20 Considering that variables in study were non-parametric, Mann-Whitney U test and Spearman' s correlation and Kruskal Wallis test were used.

Results

We found that there are differences between CHD patients and the control group when it comes to their performance in neuropsychological tests. This evaluation revealed that CHD patients had lower scores, when compared to the control group, in such tests like the Wechler's Digit Test, in direct (u=219.000; p<0,001) and in the indirect (u=«134.500; p<0.001); Rey's Complex Figure, copy (u=149.000; p<0,001) and memory (u=134.000; p<0.001); Wechler's Symbol Search (u=778.000; p=0,016); Key Search Test (u= 153.500; p<0.001); Stroop Test, words (u=131.000; p<0.001), colors (u=211.500; p<0.001) and interference (u=123.000; p<0.001); Trail Making Test, A (u=1.033.000; p<0.001) and B (u=1.000.000; p<0.001).

When analyzing type of CHD, cyanotic and acyanotic, we came to the conclusion that cyanotic patients had lower scores on the tasks, such as Wechler's Digit Test, in direct (u=803.000; p=0,018) and in the indirect (u=768.500; p=0.048); Rey's Complex Figure memory (u=848.500; p=0.004); Stroop Test, words (u=775.500; p=0.043), colors (u=779.500; p=0.038) and interference (u=819.000; p=0.011); Trail Making Test, A (u=433.000; p=0.042) and B (u=413.500; p=0.024) and Wechler's Logical Memory (u=782.500; p=0.035).

When analyzing if there are any differences between the groups divided by cardiac disease, TF, CIV and TGA, Kruskal Wallis test was used for statistical analysis. The tests in which differences were found where Direct Digits, Indirect Digits, Rey's Complex Figure – memory, Stroop Test – words, colors and Interference, Trail Making Test A and B and Logical Memory. Between patients with CIV and TF, no differences were found. When it comes to performance in these tests between CIV and TGA, we were foud differences in Rey's Complex Figure – memory (u=150.000; p=0.004), Stroop Test – Words (u=170.500; p=0.015), Stroop Test – Interference (u=160.000; p=0.008) and Trail Making Test A (u=193.000; p=0.048). No differences were found when comparing TGA and TF performances in the tasks mentioned above.

In order to verify the existence of a correlation between neonatal variables and cognitive performance in the neuropsychological tasks, Spearman's ordinal correlation was used.

We can say that in CHD patients group, neonatal variables are positively correlated. Many correlations can be found in the following variables, head circumference in relation to:

- Direct Digits (ρ=0.361; p= 0.011);
- Inversed Digits (ρ=0.294; p=0.041);

- Rey's Complex Figure copy (ρ=0.322; p=0.024);
- Key Search Test (ρ=0.351; p=0.013);
- Stroop Tests words (ρ=0.316; p=0.027);
- Stroop Test colors (ρ=0.344; p=0.015);
- Stroop Test interference (ρ=0.366; p=0.010);

Was also found a negative correlation between head circumference in relation to Trail Making Test, part A (ρ=-0.403; p=0.004). The Apgar Score 2 showed a positive correlation with the Stroop Test words (ρ=0.264; p=0.047). At last, another positive correlations can be found between weight and Rey's Complex Figure copy (ρ=0.287; p=0.023) and Key Search Test (ρ=0.367; p=0.003).

When analyzing CIV patients, many positive correlations can be found between:

- head circumference and Key Search test (ρ=0.574; p= 0.003);
- length and Stroop – words (ρ=0.406; p= 0.040);
- weight and Key Search test (ρ=0.493; p= 0.009);

As for TF patients, positive correlations were found between:

- Head circumference and Direct Digits (ρ=0.602; p= 0.029);
- Head circumference and Indirect Digits (ρ=0.579; p= 0.038);
- Head circumference and Stroop Test – Interference (ρ=0.672; p= 0.012);
- Apgar Score 2 and Stroop Test – words (ρ=0.511; p= 0.043);
- Apgar Score 2 and Stroop Test – color (ρ=0.548; p= 0.028);
- Weight and Rey's Complex Figure copy (ρ=0.570; p=0.009).

Finally, when it comes to TGA patients, positive correlations were found between:

- Apgar Score 1 and Trail Making Test part B (ρ=0.526; p= 0.044);
- Apgar score 2 and Trail Making Test part A (ρ=0.533; p= 0.041);
- Apgar score 2 and Trail Making Test part B (ρ=0.560; p= 0.030).

We also found negative correlations in the group mentioned above, as differences were found between:

- Weight and Rey's Complex Figure memory (ρ=-0.507; p=0.045);
- Weight and Stroop Test – Interference (ρ=-0.501; p= 0.048).

Discussion

According to the results, it is possible to confirm one of our hypothesis, saying that adolescents with congenital heart disease, when compared to the control group, have

lower performance in neuropsychological tasks in almost every dimension studied with the exception of Wechler's Logical Memory. What it is commonly found in current publications is that children with congenital heart disease, in fact show lower performance in neuropsychological evaluation, in several cognitive domains.[4]

Congenital heart disease children had more difficulties in auditory-verbal immediate attention tasks (Direct Digits), selective attention (Stroop Test Word, Color and Interference) and divided attention (Trail Making Test – part B). This may be explained by previous studies were congenital heart disease patients had lower scores on tasks related with attention domains[4,5,6,7] and by results of behavioural assessment evaluation by the parents of children with congenital heart disease, having described attention problems.[24]

In tasks as psychomotor ability, speed of execution and perceptive organization (in Wechler's Symbol Search), children with CHD show lower performance when compared with healthy children. This fact is corroborated by other studies[2].

As for Key Search Test, we came to the conclusion that congenital heart disease children have poorer performance. This may suggest some difficulties in executive functions, such as planning ability. Our results go along other studies that state the existence of alterations on executive functioning, planning ability, organization and problem resolution.[25,4,5,26] Alterations on executive functioning are also consistent with low results in Trail Making Test – part B.

In this study, we were able to verify that congenital heart disease patients show deficits in visual-constructive ability Rey's Complex Figure – copy and visual-spacial (Rey's Complex Figure - Memory e Trail Making Test – part A). According to Miatton et al[4], Brosig et al[6]; Bellinger et al[2]; Bellinger et al[25], children with different types of congenital heart disease reveal difficulties in tasks involving visual-spacial abilities. Visual-spacial deficits may be implicated in perceptive organization.[2]

We also found that congenital heart disease patients have worse performance in visual memory tasks (Rey's Complex Figure - memory) and working memory (Inverse Digits).

Other studies also show that these children have memory deficits Bellinger et al[2]; Miatton et al[4]; Majnemer et al[26], more specifically in working memory[2] and visual memory.[28] According to Tindall et al[28] children with congenital heart disease do not show any alterations in their verbal memory, going towards our findings. In the mean time, other studies claim that congenital heart disease children do not have any memory deficits, when compared to healthy children.[29]

In our study comparisons between neuropsychological tasks performance and children with cyanotic and acyanotic type of congenital heart disease was also made.

When assessing tasks such as Stroop Test Words, Stroop Test Color and Stroop Test Interference, , selective attention, we found that children with cyanotic congenital heart disease have worse results, when compared with children with acyanotic congenital heart disease, which suggests that cyanotic children have more difficulties when it comes to visual memory and selective attention. Hovels-Gurich et al[30] believed that children with cyanotic congenital heart disease have higher risk of developing attention problems, when compared with children with acyanotic congenital heart disease.

Children with cyanotic congenital heart disease also have worse results in Direct and Indirect Digits, Rey's Complex Figure – memory, Trail Making Test (A and B) and Wechler's Logical Memory. This fact suggest that children show more difficult in abilities such as auditory-verbal immediate attention, memory work, visual constructional memory, visual-spacial orientation, divided attention, executive functioning and immediate verbal memory, when compared with children with acyanotic congenital heart disease.

Even though differences were found in neuropsychological performance between cyanotic and acyanotic groups, this data are not completely consistent with other studies, where there are no differences in neuropsychological performance between these groups.[27,31]

In order to further understand in which sub-groups, CIV, TF and TGA, there are differences in Direct Digits, Indirect Digits, Rey's Complex Figure – memory, Stroop Test – words, colors and Interference, Trail Making Test A and B and Logical Memory, an analysis was made between CIV and TF; CIV and TGA; and TF and TGA.

In the first group, CIV and TF, no differences were found. When it comes to the second group, CIV and TGA, we had differences in performances in Rey's Complex Figure – memory, Stroop Test – Words, Stroop Test – Interference and Trail Making Test A.

No differences were found between TGA and TF in these tests performances.

As for neonatal variables, as head circumference, weight and Apgar score (first and fifth minute) this may show vulnerability of mental development and psicomotor.[23] In this way it is important to study the existing correlations between neonatal variables and cognitive performance in neuropsychological tasks in adolescents with congenital heart disease.

When talking about the different congenital heart disease groups (CIV, TF and TGA), we can see that the head circumference is positively correlated with auditive verbal immediate attention, working memory, visual constructive ability, visual memory, planning abilities and selective attention. We also found a negative correlation in this same variable with psicomotor performance.

It seems that head circumference can be an indicator of poor performance in many cognitive tasks. Through blood flow auto-regulation, several brain areas can be protected by cerebral vasodilatation, in cases of problems with fetal oxygenation, which may be harmful, as it is in congenital heart disease.[1]

When talking about variables like Apgar score and weight, we came to the conclusion that there is a positive correlation with selective attention and with visual-constructive ability and planning ability, respectability.

With this results it is now possible to say that neonatal variables are indeed related with the ability to keep their attention in adolescents with congenital heart disease.

As for CIV subgroup, we found positive correlations between head circumference and planning ability. This data suggests that children with congenital heart disease have small head circumference, having long term consequences in their neurodevelopment.[3,1]

In this same group there are also positive correlations between length and selective attention, and weight and planning ability.

As mentioned earlier, in cases of oxygenation problems, there is a global restriction of somatic growth, due to the redistribution of cerebral circulation.[32] In this way, a delay in fetal growth may have an influence in long term neurocognitive performance.

In TF subgroup, there are positive correlations between head circumference and auditive verbal immediate attention, working memory and selective attention; Apgar score 2 and selective attention, and weight and visual constructive ability.

Congenital heart disease newborns have a tendency to have hipoxicoisquemic lesions,[33] that can be detected through magnetic resonance imaging,[34] and this can be related to the low results in Apgar score, and later on these may have repercussions on the child's cognitive performance.[32]

This group, TF, was the one where the least amount of differences were found between neonatal variables and neurocognitive performance.

The fact that the weight can only be related with neurocognitive behaviour, this does not completely agree with previous studies, as according to Miatton et al[4,] low weight in children with TF can have an influence on neurocognitive performance.

Finally, in TGA group, we found a positive correlation between Apgar scores 1 and 2 and executive functioning, such as planning ability, cognitive flexibility and altering attention. A low Apgar score, at 5 minutes, may be an indicator of neurological damage, that can be translated into long term neurocognitive deficits.[33]

As for weight, we were able to find a negative correlation with visual memory and selective attention. The last which can lead us to say that this variable may interfere with the ability to inhibit interference.

With these results we were able to verify that neonatal variables have an influence on different cognitive domains, being the most representative one the congenital heart disease group.

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Anexo 2

Co - Autora de Um Artigo submetido para British Heart Journal

PSYCHOSOCIAL ADJUSTMENT AND PRONENESS TO PSYCHOPATOLOGY IN ADOLESCENTS AND YOUNG ADULTS WITH CONGENITAL HEART DISEASE

Maria Emília Guimarães Areias,
Rua Central de Gandra, 1317,
4585-116 Gandra PRD – Portugal
T:+351 224 157 176
F:+351 224 157 102
metega@sapo.pt ; memilia.areias@cespu.pt

Isabela Freitas [1,4]

Marta Castro [1,4]

Sofia Sarmento [1,4]

Rosália Coelho [1,4]

Flávio Teixeira [1,4]

Victor Viana [2,4]

José Carlos Areias [2,5]

Maria Emília Guimarães Areias [1,3]

[1]Department of Psychology, Instituto Superior de Ciências da Saúde do Norte (CESPU), Gandra, Portugal
[2]Department of Pediatrics (Cardiology), Hospital São João, Porto Medical School, University of Porto, Portugal
[3]CINEICC, Centro de Investigação do Núcleo de Estudos e Intervenção Cognitivo-Comportamental, Coimbra, Portugal
[4]UNIPSA, Unidade de Investigação de Psicologia e Saúde/ CICS (CESPU), Gandra, Portugal
[5]Unidade de Investigação Cardiovascular da Faculdade de Medicina do Porto, Portugal

Key Words: Congenital Heart Disease, Psychosocial Adjustment, Psychopathology

ABSTRACT

Objectives: Our purpose was to study psychosocial adjustment and psychiatric morbidity of adolescents and young adults with congenital heart disease (CHD). **Methods:** We evaluated 110 CHD patients (62 male) aged from 12 to 26 years old (mean=18.00 ± 3.617), 58 cyanotic. All assessment measures were once obtained in a tertiary hospital. Demographic information and clinical history were collected. Questionnaires regarded topics as social support, family educational style, self-image and physical limitations, a standardized psychiatric interview SADS-L, and a self-report questionnaire on psychosocial adjustment, YSR or ASR. One of the relatives completed an observational version of the same questionnaires (CBCL or ABCL).

Results: We found a 21.8% lifetime prevalence of psychopathology, 31.3%, in females, 14.5% in males, showing a somewhat increased proneness in CHD patients. Females also showed worse psychosocial adjustment, with more somatic complaints (u=260.000; p=0.011), anxiety/ depression (u=984.000; p=0.002), aggressive behavior (u=920.500; p=0.001), attention problems (u=1123.500; p=0.027), thought problems (u=1069.500; p=0.010), internalization (u=869.000; p=0.000) and externalization (u=1163.000; p=0.050). Patients with severe CHD (u=939.000; p=0.030) and surgical repaired (u=719.000; p=0.037) showed worse psychosocial adjustment. Those with poor social support showed more withdrawn (u=557.500; p=0.000) and social problems (u=748.500; p=0.023), and patients with unsatisfactory school performance revealed more anxiety/depression (u=916.000; p=0.020) and attention problems (u=861.500; p=0.007).

Conclusions: CHD males with good social support and good academic performance have a better psychosocial adjustment.

Key Words: Congenital Heart Disease, Psychosocial Adjustment, Psychopathology

Abbreviations: CHD - congenital heart disease; SADS-L - Schedule for Affective Disorders and Schizophrenia – Lifetime version; YSR - Youth Self Report; CBCL - Child Behavior Checklist; ASR - Adult Self Report; ABCL - Adult Behavior Checklist

INTRODUCTION

The survival rate in the 1950's for children born with moderate CHD was about 20% whereas today about 90% of these children achieve adulthood.[1]

There has been a decrease in child mortality thanks to advances over the last four decades in diagnostic, surgical and catheter interventional techniques.[2] As these children survive, the interest in issues such as psychosocial outcomes have increased also.[3]

Most children with CHD were diagnosed in uterus or in infancy, and are expected to undergo surgical procedures either to correct or palliate their defect.[4, 5] These children need to be seen regularly by a cardiologist.[4]

Many studies have been conducted assessing the impact of CHD on children's or adolescents' psychosocial and cognitive functioning. Although, a consensus among these studies have not yet been reached, some report higher rates of behaviour problems in children and adolescents with CHD, while others have not found any differences between patients with CHD and norms.[4]

It is believed that children with CHD have a higher risk of developing behavioural and emotional problems, when compared to healthy children. Several studies have reported that these children have increased feelings of anxiety and inferiority, higher degrees of impulsiveness, higher levels of emotional and behavioural problems.[6] On the other hand, main European studies have showed a good psychological functioning in adults with CHD.[7]

Not much is known about this topic, as some studies say that, in a 25-year follow-up, more psychosocial distress was found in adults with CHD in comparison with a normative group. The differences found were limited to somatic complaints and thought problems and behaviours.[6]

As for psychopathology, studies have also disagreed in some aspects, many authors believe that CHD patients have a higher probability of having psychopathological symptoms while others have found similar numbers between these patients and healthy children and adolescents.[8]

Several characteristics can be described as facilitators of positive perspectives of stressful life situations and reduced psychological distress such as self-esteem and similar conduct like self-concept and self-perception. Some studies found that usually CHD patients have lower self-esteem although, after surgery, patients reported better self-esteem or self-concept.[9]

Cognitive perceptions are believed to have an influence on a CHD patient's life. The more negative these perceptions are, the higher psychological distress was found. The negative perception can be associated, more than the severity of the disease itself, to higher distress and worse psychological adjustment.[9]

Some studies have shown that patients with cyanotic heart disease have a higher risk of presenting behavioral problems compared to patients with non-cyanotic heart defects, but other studies did not show this association.[5, 10]

Patients with CHD who underwent surgical procedures, had more behavioral problems when compared with those who did not require surgery, and more likely to develop psychiatric problems.[5]

As far as the physical condition is concerned, most patients with CHD have limitations, that leading to more behavioral and emotional problems.[5, 8]

In this study, we aimed to evaluate psychosocial adjustment and proneness to psychopathology in adolescents and young adults with CHD. The importance of our investigation is that it systematically addressed the question of how the several demographic and clinical variables relate to psychiatric morbidity and to psychosocial adjustment, using very strict methods of psychiatric diagnosis.

METHODS

Participants

The study enrolled 110 CHD patients, 62 male and 48 female, with a mean age of 18.00 ± 3.62 years (range:12-26 years old). The participants who had not achieved an educational level that enabled them to understand and complete the written questionnaires were excluded from the study.

At the time of the interview, two participants were married, one was divorced, two were living in a marital union. All the others (105) were single.

53 patients had completed their secondary education (12^{th} grade), 40 the 3rd cycle (9^{th} grade), 11 the 2nd cycle (6^{th} grade) and 6 had graduated from college. Of these patients 55 had at least repeated one year at school (mean=1,49 ± 0,50 year).

Of the 110 participants, 20 were employed full- or part-time, 7 were unemployed and all the others 83 were students.

Complete medical records were available for all the patients, who had been followed in the pediatric cardiology or cardiology departments of a tertiary hospital.

For 58 individuals the CHD was cyanotic and for 52 it was acyanotic; 34 of these patients had a severe form of CHD, 18 a moderate and 58 a mild one; 41 patients had some physical limitations while 69 did not. 4 patients had severe residual lesions, 21 moderate and 85 mild lesions. 23 patients were never submitted to any kind of surgical procedure, while 42 had 1 surgery, 25 had two, 11 had three, 5 had four, 3 had five and 1 had 9 surgeries. 47 patients were on pharmacological therapy while 63 were not.

In many of our participants, the main CHD was combined with other heart diseases. Patients with associated cardiac malformations or chromossomopathies were excluded from the study. The participants had the following distribution of pathologies: transposition of the great arteries (9; two of them had also ventricular septal defect and aortic stenosis, and one had ventricular septal defect and pulmonary stenosis), tetralogy of Fallot (30), coarctation of the aorta (11; one had also ventricular septal defect and one aortic stenosis) ventricular septal defect (24; one had also interruption of the aortic arch and one had mitral insuficiency), atrial septal defect (6; one had also mitral atresia and pulmonary hypertension, and one had Ebstein disease), atrioventricular septal defect (4), aortic stenosis (6), pulmonary stenosis (6), single ventricle (2; one had also pulmonary atresia and one had pulmonary stenosis), patent ductus arteriosus (2), double-outlet right ventricle (1), pulmonary atresia (3), Ebstein disease (3), mitral valve prolapse (1), bicuspid aortic valve (1) and tricuspid valve regurgitation (1).

The diagnosis was determined during the neonatal period for 61, before the first birthday for 28, 5 were diagnosed between the ages of 1 to 3 years, 6 were diagnosed between the ages of 3 to 6 years and between the ages of 6 and 12 for 12 participants.

The first surgery was performed for 5 of the participants during the neonatal period, before the first birthday for 30, between the ages of 1 and 3 for 19, and between the ages 3 and 6 for 20 participants, between the ages 6 to 12 for 8 and between the ages of 12 to 18 for 88.

We also invited one relative of each patient to participate in this study and 100 accepted to take part in it.

Assessment Instruments

In this study, we used different surveys to collect the necessary information: an identification form, a semi-structured interview, a standardized psychiatric interview SADS-L, the self-report and observational questionnaires of the ASEBA system for psychosocial adjustment, ASR, YSR, ABCL and CBCL (for patients > 18 and < 18).

Additional questionnaires used in this research are described in detail in another report. We used an identification form to collect personal and demographic data from each patient (e.g., marital status, educational level and occupation), as well as all relevant aspects from their medical history (diagnosis, severity and category of heart disease, surgical interventions, pharmacological therapy, and presence of residual lesions, among others).

The semi-structured interview included 38 multiple-choice or short-answer questions that focused on different topics such as social support, family upbringing, self-image, functional limitations and emotional adjustment.

A standardized psychiatric interview, SADS-L (Schedule for Affective Disorders and Schizophrenia – Lifetime version) [11], was administered to obtain a clinical diagnosis of any psychopathological disorders that may have existed before the interview in these patients.

The YSR and ASR are self-report questionnaires, designed to collect a description of a child or adult's functioning; they assess individuals in scales of withdrawn behavior, somatic complaints, anxiety/depression, thought problems, social problems, attention problems, delinquent behavior, aggressive behavior, internalization and externalization.[12] The CBCL and ABCL are observational versions of the same questionnaires, to be completed by the patients' parents or caregivers, having as a requirement being knowledgeable about the patient, as they report their perception on the behavior and possible problems occurring in the patient. For their similarities, and to have a better representative sample, the results of the YSR and ASR were pooled, as well as the results of the CBCL and the ABCL, and for statistical purposes the overall results were counted for each scale.

Procedure

Prospective participants were contacted while waiting for their appointment in cardiology or pediatric cardiology department. At this time, they were informed about all aspects of the research, and when they accepted to participate, they were asked to sign a consent (either the patients themselves or the caregivers when they were under 18 years old). The interview happened on the spot. The parents or caregivers accompanying the patient were asked to fill out a questionnaire, and 10 caregivers refused to participate or were not present for the application of the protocol, and subsequently expressed their intention not to participate.

Design

All the assessment measures were obtained on a single occasion. Clinical data were collected retrospectively using each patient's clinical record, with assistance from hospital medical staff.

Data Analysis

Statistical analysis of the data was processed using the software IBM SPSS (Statistical Package for the Social Sciences, Chicago, IL, USA), version 19. The distribution of all the variables was tested. Differences for parametric variables were established using Student's t-tests, while differences for non-parametric variables (the majority) were established using Mann-Whitney U test and Chi-square tests of association.

RESULTS

We found that 21.8% of our participants had a psychiatric disorder and that there was a statistical difference between the two genders, with females almost doubling males' rate (31.3% in females and 14.5% in males; p=0.035). One or more of the following psychiatric disorders had been diagnosed for our participants in all their lifetime prior to interview: Minor or Major Depressive Syndrome (13), Panic Disorder (3), Anxiety Disorder (4), or Manic Syndrome (3), Cyclothymic Personality (1).

For the sake of data analysis, we grouped the results on psychosocial adjustment in either self-reported or observational.

The self-report measures on psychosocial adjustment revealed statistical differences between genders, as females showed higher levels of somatic complaints (u=260.000; p=0.011), anxiety/ depression (u=984.000; p=0.002), aggressive behavior (u=920.500; p=0.001), attention problems (u=1123.500; p=0.027), thought problems (u=1069.500; p=0.010), internalization (u=869.000; p=0.000) and externalization (u=1163.000; p=0.050) and overall worse psychosocial adjustment than males. On the other hand, teens and young adults with a severe CHD showed worse psychosocial adjustment, with more somatic complaints (u=264.000; p=0.022) and higher levels of internalization (u=917.000; p=0.015) in self-report measures, when compared with patients with moderate-to-mild CHD.

When analyzing the impact of the kind of CHD in psychosocial adjustment, no statistical differences were found.

Patients who underwent surgical interventions revealed worse psychosocial adjustment than patients with no surgical repairs, showing higher levels of withdrawn behavior (u=719.500; p=0.037) in self-assessment.

Patients with poor social support reported worse psychosocial adjustment, with higher levels of withdrawn (u=557.500; p=0.000) and social problems (u=748.500; p=0.023) when compared with patients with good social support.

Patients with limited physical competence showed more withdrawn behavior (u=1023.000; p=0.015) when compared to patients with satisfactory physical competence, thus presenting worse adjustment.

Patients with worse academic performance showed higher levels of anxiety/depression (u=916.000; p=0.020) and attention problems (u=861.500; p=0.007) in self-report, when compared to those who feel their academic performance was satisfactory.

Patients with severe-to-moderate residual lesions revealed worse psychosocial adjustment in self-report than those with mild residual lesions, showing higher levels of internalization (u=782.500; p=0.046).

No differences were found in self-report between patients with or without need for pharmacological therapy.

According to the assessment of patients' caregivers, no differences were found between patients who underwent surgical procedures and the ones who did not had surgical procedures done.

In the caregivers' assessment, male patients are perceived as having worse psychosocial adjustment than females, as they were assessed as having higher levels of withdrawn (u=911.500; p=0.020) and aggressive behaviors (u=945.500; p=0.038).

Again on the relatives' standpoint, the cyanotic patients were assessed as having higher levels of attention problems (u=981.500; p=0.045) than the acyanotic, thus showing worse psychosocial adjustment.

When compared with patients with good social support, those with poor support showed, also on the stand point of the caregivers, higher levels of withdrawn (u= 517.000; p=0.001) and internalization (u=608.000; p=0.007) and thus, worse psychosocial adjustment. According to their relatives' assessment, patients with severe to moderate forms of residual lesions showed higher levels of social problems (u=205.500; p=0.008), attention problems (u=649.500; p=0.028) and internalization (u=567.500; p=0.004), than the ones with moderate to mild residual lesions, and thus a worse psychosocial adjustment.

DISCUSSION

This study is important because it systematically tested the effects of different demographic, clinical and psychosocial variables in psychosocial adjustment and psychiatric morbidity. In a growing population of adults with CHD, this information is rather important in unveiling strategies that can be used to assist and care for patients, leading to better emotional adjustment and better outcomes in different life challenges.

The assessment instruments used for psychosocial adjustment enabled us to compare sub-groups of patients, regarding demographic, clinical and psychosocial variables. One main finding of our study was that females with CHD reported higher levels of somatic complaints, anxiety/depression, thought problems, attention problems, aggressive behaviour, internalization and externalization, than males, thus showing a worse psychosocial adjustment. Contrariwise, relatives find male patients more withdrawn than girls.

In our study, female patients had almost the double of the lifetime prevalence of psychopathology than males. These findings on psychiatric morbidity and on the different scales of psychosocial adjustment are consistent with other studies on the general population that report differences between genders, with females showing higher rates of emotional problems. Studies show also that females have greater likelihood of displaying higher levels of anxiety/depression

and somatic complaints when facing negative obstacles that interfere with the interpersonal level, resulting in higher levels of internalization.[6,8,13]

This may be due to the presence of a scar, situated on the chest, being a source of uncertainties or discomfort. In addition to affecting sexual relationships CHD can also interfere with pregnancy and delivery, leading to a sense of anxiety about their physical condition.[6,8]

Some studies show that females are more likely to develop depressive symptoms when facing negative life events than males.[6,10]

In this study, adolescents or young adults with severe type of CHD reported having higher levels of social problems and, thus, worse psychosocial adjustment, compared with those with moderate or mild form of CHD.

These results may be related to the fact that they need further medical care throughout their life, while patients with mild or moderate CHD may have a daily life similar to healthy adolescents and young adults.[8] Patients with severe forms of CHD show higher level of internalization and somatic complaints and that may be associated to the fact that these patients are more vigilant about their health, being more anxious about any complications. This may explain the results, since anxiety is a component of internalization scale.[14]

The type of CHD did not show any impact with statistical relevance in patients' selfreport measures of psychosocial adjustment. However, the caregivers' standpoint seems to be more sensitive regarding this feature, as they perceive the cyanotic patients as having more attention problems and worse psychosocial adjustment.

Other published studies also showed that the cyanosis is not a stable indicator that patients will have behavioural and emotional problems.[8,10,15]

Patients who underwent surgical procedures revealed higher levels of withdrawn behavior. This may be related with the fact that admissions are long as well as the recovery, thus providing a prolonged absence from education and from contact with the peer groups, which could lead to difficulties of reintegration and therefore to the isolation of patients.[5,7,16]

Patients with worse social support had higher levels of withdrawn behavior and social problems, and thus, a worse psychosocial adjustment. The assessment of the caregivers also reported higher levels of withdrawn behavior and internalization in patients with poor social support, showing worse psychosocial adjustment.

According to several studies, parents and siblings of adolescents or young adults with CHD are more prone to face a different number of psychosocial stresses putting the whole family in need of psychosocial support. Many studies reveal a higher need for intervention on family problems in families with children with chronic medical diseases. When the complexity of the disease is low, parents seem to be more fitted to provide support.[17] These families are reported to experience more stress, that can have an impact on the child's adjustment.[15]

Parents of children with CHD can be overprotective and hypervigilant about their child's health, making it hard for their children to be more independent. Many studies show that these patients are more likely to have "dependent lifestyles" than healthy adolescents or young adults.[7] The participation in leisure time activities can be a contributor to a better social outcome.[13]

Limited physical competence translated into more withdrawn, feeling more isolated, when compared with patients with satisfactory physical competence. Self-report showed that patients with physical limitations have worse psychosocial adjustment. A low exercise capacity can be translated into more internalizing problems. For older heart patients, limited physical competence lead to concerns and anxiety about their health.

According to some authors, patients submitted to physical training interventions, showed a decrease in internalizing problems.[8]

Physical limitations and school absences prevent full participation in different activities, leading to isolation and social awkwardness. This can be translated into restricted employment opportunities.[7]

In our study, an unsatisfactory academic performance led to worse psychosocial adjustment, as patients report having higher levels of anxiety/depression, attention problems and externalization than those with good academic performance. Several previous published studies show that CHD has an impact on school careers, for the many hospitalizations and restrictions, being the main reason for the attendance of special education by these patients. When compared to healthy adolescents or young adults, the CHD patients are more unlikely to complete a lower educational level.[13]

Sometimes, children with CHD have neurodevelopment deficits. These often will not show until school age, when the academic demands start having an impact on their lives. Many families rationalize their child's developmental delay to the disease and the several hospitalizations.[18]

Some studies show that unsatisfactory educational background can be translated into lower educational and occupational achievement.[7]

This study showed a 21.8% prevalence of psychiatric disorder in our patients. Females showed a higher percentage of psychiatric disorder with 31%, and males only had 14%. When compared to the reference value of the World Health Organization (WHO), 10% of the world population, it seems that adolescents and young adults with CHD have an

increased proneness for psychiatric diagnosis.[19] However, a study of six different European countries showed a prevalence of 25% in the general population, which is closer but higher than the results for CHD patients in our study.[20] Another study estimated that the life time prevalence of psychopathology is 19.4% in Spain, 18.1% in Italy (countries that can be considered culturally close to Portugal), and 25.2% in Germany, but in striking contrast, 37.9% in France and 47.4% in the United States of America.[21]

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COMPETING INTERESTS

There are no competing interests.

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Anexo 3

Co-Autora de Um Artigo pronto a submeter para British Heart Journal

ADOLESCENTS AND YOUNG ADULTS WITH CONGENITAL HEART DISEASE: QUALITY OF LIFE

Maria Emília Guimarães Areias,
Rua Central de Gandra, nº 1317,
4585- 116 Gandra PRD – Portugal
T:+351 224 157 176
F:+351 224 157 102
metega@netcabo.pt ; memilia.areias@cespu.pt

Marta Castro [1,4]

Isabela Freitas [1,4]

Sofia Sarmento [1,4]

Rosália Coelho [1,4]

Flávio Teixeira [1,4]

Ana Margarida Silva [1,4]

Cláudia Vaz [1,4]

Daniela Vieira [1,4]

Cidália Proença [1,4]

Cláudia Moura [2,5]

Victor Viana [2,4]

José Carlos Areias [2,5]

Maria Emília Guimarães Areias [1,3]

[1]Department of Psychology, Instituto Superior de Ciências da Saúde do Norte (CESPU), Gandra, Portugal
[2]Department of Pediatrics (Cardiology), Hospital São João, Porto Medical School, University of Porto, Portugal
[3]CINEIC, Centro de Investigação do Núcleo de Estudos e Intervenção Cognitivo-Comportamental, Porto, Portugal
[4]UNIPSA, Unidade de Investigação de Psicologia e Saúde, Gandra, Portugal
[5]Unidade de Investigação Cardiovascular, Porto, Portugal

Key Words: Congenital Heart Disease, Quality of Life

Abstract

Aims: The objective of this essay is to assess quality of life (QoL) within a population of teenagers and young adults with congenital heart diseases (CHD).

Methods: 110 patients with CHD participated in this study, 62 male and 48 female, aged 12 to 26 (average= 18,00 ± 3,617). It was taken only once and relevant aspects, such as clinical and demographic data, were collected. Furthermore, a set of psychological evaluation instruments was applied, constituted by a semi-structured interview, a standard psychiatric interview (SADS-L) and a questionnaire on quality of life (QOL-BREF).

Results: Compared to the general Portuguese population with no historical data of the disease, our sample has shown a better QoL in every domain, except for the general quality of life domain, revealed by the results of physical (t = -12,277; p = 0,000), psychological (t = -2,524; p = 0,013), social relationships (t = 3,566; p = 0,001) and environment (t = 5,776; p = 0,000) domains.

Contrarily, statistically significant differences were not found as far as the psychiatric diagnosis and the severity of CHD are concerned.

Contrasting patients submitted to surgical interventions, a set of patients without surgeries demonstrated a better quality of life, as we can see through the physical (t=-2,525; p=0,013), psychological (t=-2,394; p=0,018), social relationships (t=-2,502; p=0,014) and general QdV (u=1294,000; p=0,006) results.

The social support has revealed fairly significant results, and appears to be an important aspect in all domains of these patients' lives - physically (t=2,707; p=0,008), psychologically (t=2,755; p=0,007), socially (t=4,976; p=0,000), environmentally (t=3,085; p=0,003) and in general QdV (u=623,500; p=0,000).

Satisfactory schooling results are also linked to QoL in the psychological (t=2,457; p=0,016), environmental (t=2,346; p=0,021) and general QdV (u=926,500; p=0,013) domains.

Conclusions: Congenital heart diseases patients show a better quality of life when they are not submitted to surgical interventions, when they have a good social support, as well as a satisfactory schooling experience.

Key Words: Congenital Heart Diseases, quality of life; social support.

INTRODUCTION

The purpose of this research was to evaluate quality of life in children and adolescents with congenital heart disease.

Congenital heart defects are the second leading cause of death in childhood and adolescence, which corresponds to approximately 1% of live births, as well as the only cause of heart disease in the pediatric population in developed countries.

Mortality in children with congenital heart disease has decreased significantly over the past four decades as a result of important advances in diagnosis, surgery and catheter intervention techniques. The age for surgical intervention has decreased substantially, by various conditions. New techniques, including catheter intervention, have been introduced to severe heart disease. [1] Thus, because of these new techniques about 85 to 90% of children with congenital heart disease can live to adulthood with a better quality of life. [2, 3] However, survival is not always a synonym of high quality of life, because these patients often face physical, psychological or cognitive difficulties. [3]

In addition, and because it is a chronic disease, these patients find themselves faced with some events triggered by the disease, particularly during school breaks, restrictions on the physical level and even restrictions that can affect their adult lives. [4]

Thus, the quality of life has become a very important factor for these patients. [1]

The assessment of quality of life should be made according to the children development stage and their level of growth, the severity of the disease, their family, the acceptance of the illness and personality traits. Furthermore, the evaluation of quality of life of children with congenital heart disease should be continuous, since the surgery can ensure survival, even though it does not allow a "normal" life, as in the postoperative period frequent disease revaluations are necessary and planned, as well as the implementation of treatment and the compliance with it.

Children with congenital heart disease experience feelings of anxiety and depression due to the frequency of hospitalizations, daily medication and the limitations imposed by the disease. The poor quality of life is usually attributed to the lack of social acceptance, especially in the school context. The disability of these patients makes them unable to fulfil their duties. Very often they have to give up activities they normally enjoy, triggering feelings of loneliness, rejection, social isolation that make social integration more difficult. School performance is often affected and children typically fall behind their healthy peers, because they have a long treatment process that involves frequent hospital admissions and prolonged absences from school. [3]

In order to overcome some of these feelings and limitations, these patients develop coping strategies that reveal very important and useful. One of the strategies children use is not thinking about the heart disease. Another strategy is related to the fact that, knowing the symptoms associated with heart disease and because these patients already know how to deal with it, in case of new symptoms, they may feel they soon will be vanished. [5]

Another strategy refers to the knowledge of their own limits, that is, patients are aware of what they are capable of, as well as situations that can cause them concern, then, they tend to avoid them. [6]

METHODS

Participants

110 congenital heart disease patients participated in the present study and were interviewed in the cardiology and pediatric cardiology services at Hospital de São João in Oporto.

We had a total of 110 CHD patients, 62 male and 48 female, aged from 12 to 26 years old (mean=18,00 ± 3,617). Regarding cardiac malformation, 58 subjects were cyanotic and 52 non-cyanotic; 34 of these patients had a severe form of CHD, 18 moderate and 58 mild; 41 patients had some form of physical limitations while 69 did not. According to their medical records, 4 patients have severe residual lesions, 21 moderate residual lesions and 85 mild residual lesions. As for surgical interventions, 23 patients have never been submitted to any kind of surgical procedure, while 42 had 1 surgery, 25 had two, 11 had three, 5 had four, 3 had five and 1 had 9 surgeries. At that time, 47 patients were on pharmacological therapy while 63 were not.

Regarding their age at first surgery, 13 participants had their first surgery in the neonatal period, 23 during their first year of life, 18 between one year and three years of life, 20 between three and six years, eight between six and twelve years and, finally, six participants had their first surgery between twelve and eighteen.

Concerning the educational background, a total of 53 patients had completed their secondary education (until the 12th grade), 40 had completed the 9th grade, 11 had completed the 6th grade and 6 had graduated from college education. 55 of these patients had at least repeated one year of school.

As for their marital status, 105 were single, 2 were married, 2 cohabited and 1 was divorced.

When it comes to the psychiatric diagnosis, 22 patients indicated symptoms of a psychiatric disorder and 88 did not. From those who were diagnosed with one or more psychiatric disorders in all their lifetime 13 presented Minor or Major Depressive Syndrome, 3 Panic Disorder, 2 Anxiety Disorder, 3 or Manic Syndrome and 1 Cyclothymic Personality.

Instruments

In order to collect all the necessary information for this study, different tools were used, such as an identification form, a semi-structured interview, a standardized psychiatric interview SADS-L ("Schedule for Affective Disorders and Schizophrenia – Lifetime version") and WHOQOL-BREF.

When approaching patients, our first concern was to get their or their parents' consent. After signing this form, an identification form followed, so that we could collect personal data from each patient, as well as aspects from their medical history such as diagnosis, type of heart disease, surgical interventions, pharmacological therapy and residual lesions, among others.

The semi-structured interviewed focused on different topics such social support, family upbringing, self-image, functional limitations and emotional adjustment; this was also intended to collect some socio-demographic data.

A psychiatric interview, SADS-L, was used to evaluate psychopathology in these patients.

The WHOQOL-BREF is a self-report questionnaire that assesses subjective QOL in both healthy individuals and those with range of psychological and physical disorders. It is a 26 item Likert-type scale with ratings from 1 to 5. For almost all the scale items, higher scores reflect a higher QOL. However, for three items (questions 3, 4 and 26), higher scores reflect a lower QOL. The first two questions assess general QOL. The WHOQOL-BREF also assesses four dimensions of QOL: physical (questions 3, 4, 10, 15, 16, 17 and 18), psychological (questions 5, 6, 7, 11, 19 and 26), social (questions 20, 21 and 22) and environmental (questions 8, 9, 12, 13, 14, 23, 24 and 25).

Procedure

The patients were invited to participate in this study while they were waiting for their cardiology or pediatric cardiology appointment. At this time, the patients themselves or, when under 18 years old, their caregivers were informed about all aspects of the research. Those who agreed completed a consent form. To make this survey possible, we have also needed to ask the Hospital Ethical Committee for permission, which followed international conventions, guaranteeing the rights of the patients. The interview happened on the spot.

Design

All assessment measures were obtained on a single occasion.

Data Analysis

The data analysis of the different instruments was processed using the software IBM SPSS (Statistical Package for the Social Sciences) Statistics, version 19. Considering that variables in this study were non-parametric, Mann-Whitney U test and Chi-Square were used.

RESULTS

Compared to the Portuguese population [7], participants in our sample indicate a better perception of quality of life in all areas, except in the field of overall quality of life. Quality of life revealed significant in the physical (t=-12,277; p=0,000), psychological (t=-2,524; p=0,013), social relationship (t=3,566; p=0,001) and environment (t=5,776; p=0,000) domains.

The severity of heart disease is complex and can be divided into mild or moderate severity. When comparing these subgroups, we find that there are no statistically significant differences between them.

With regard to residual lesions, there are only statistically significant differences in the physical quality of life (t=- 2,432; p=0,017).

When comparing patients who need treatments using medication with patients without medication, these last ones had better QOL in the area of social relationship (t=- 2,091; p=0,039).

The patients that were not submitted to surgical interventions showed better QOL in all areas, except in the area of environment – Physical (t=- 2,525; p=0,013), Psychological (t=- 2,394; p=0,018), Social Relationship (t=- 2,502; p=0,014) and general QOL (u=1294,000; p=0,006). Furthermore, patients with less than two surgeries have higher levels of quality of life in all dimensions, except in social relationship domain – Physical (t=- 2,469; p=0,015), Psychological (t=- 1,879; p=0,047), Environment (t=- 2,547; p=0,012) and general QOL (u=443,000; p=0,025).

Social support revealed a great impact on QOL. The patients with better social support had better QOL in general and across all dimensions – Physical (t=2,707; p=0,008), psychological (t=2,755; p=0,007), Social Relationship (t=4,976; t=0,000), environment (t=3,085; p=0,003) and general QOL (u=623,500; p=0,000). Furthermore, the female participants reported greater social support than the male participants ($\chi^{2=}$ 7,349; p=0,011).

Patients with satisfactory physical competence showed better QOL when compared with patients with limited physical competence in the physical (t=- 2,093; p=0,039), psychological (t=- 2,674; p=0,009) and general QOL (u=971,500; p=0,002) dimensions.

Regarding the patients with satisfactory educational background, they had better QOL when compared with patients with an unsatisfactory educational background in three domains,

specifically, in the psychological (t=2,457; p=0,016), environment (t=2,346; p=0,021) and general QOL (u=926,500; p=0,013) domains.

Acyanotic patients showed better QOL comparatively to cyanotic patients in the physical domain (t=- 2,196; p=0,033).

When comparing the quality of life of the age groups, 12 -18 and 19 -26, there were only statistically significant differences in the psychological domain (t=-2,466; p=0,016).
Finally, comparing the gender with the various domains of quality of life, there were significant differences in the field of the environment (t=2,258; p= 0,026).

DISCUSSION

The given results made possible the analysis and comparison with previous researches. A range of variables – age, gender, submission to surgical interventions, number of surgeries, presence of cyanosis, social support, schooling process, physical ability, residual lesions, presence or absence of psychopathological disorders and pharmacological therapy – were tested to check their influence on the quality of life of these patients.

When compared with the overall Portuguese population, patients with congenital heart disease showed a better quality of life in all the domains, except for the general quality of life.

Unlike the previous study, there is another piece of research in which the quality of life of these patients and the global population is assessed; the results are similar to those of our research, showing that patients with congenital heart disease have a better quality of life than the general population. [8, 9] According to some authors, it can be explained by the fact that patients with congenital heart disease accept their health condition and their restrictions. [10]

In what is related to the presence or absence of cyanosis, only significant statistically differences were found in the physical domain in patients with acyanotic CC type, revealing a better quality of life than those of patients with cyanosis. These results can be proved with other studies by Landolt, Buechel & Latal (2008), since they confirm that the presence of cyanosis can influence the quality of life. [11]

Regarding the submission to surgeries, patients who were not submitted to surgeries have better results in all the domains, except for the environment, when compared with patients who were not. In fact, there are some explanations for this – lots of patients submitted to surgeries have body scars because of surgical corrections and, in spite of developing many life skills, the severity of the disease always end up conditioning their lives. Besides, the need of surgical corrections during their lifetime often interferes with their future planning. [12]

Concerning the severity of the heart disease, the results did not show any statistically significant number in any of the approached domains, what suggests that this factor does not influence the quality of life of the patient. The same fact was also confirmed by the conclusions of Ternestedt et al. (2001) and Krol et al. (2003). [13, 6]

Statistically significant results related to residual lesions were disclosed only in the physical domain – patients with mild residual lesions tend to show a better quality of life than patients with severe residual lesions. This matches the results from another study that proves that the existence of severe residual lesions and a more unstable clinical frame predicts a worse quality of life. [10]

With reference to the pharmacological therapy, through the results, we could notice a better quality of life only in the social relationship domain when patients did not have to accede to it. According to Spijkerboer and his collaborators, the pharmacological therapy is associated with the difficulties in the emotional field as well as of behaviour, and consequently, with cases of severe heart disease. [14] Another fact showed that, due to pharmacological therapy and regular hospital admissions, patients often develop feelings of anxiety and depression. [3]

On the topic of the psychiatric diagnosis, statistically significant differences were not found in any of the domains of quality of life, which possibly means that this is not a quality of life conditioning factor. Another piece of work confirms the same – though the frequent hospital admissions, even at early ages, linked to other problems in the emotional and behavioural levels, there are other factors – the fact of being the first surgery or the post-surgery period do not seem to be connected to the presence of psychopathology. [15]

Very significant conclusions were revealed by the social results, making clear that this is a very relevant factor in the patients' lives, in all the domains – physical, psychological, social relationships, environment and the general quality of life. As Tak & McCubbin sustained (2002), the social support is a primary aspect either in the individual or in the familiar field, as well as within the patients' community, and plays a crucial role in the balance between stress and

psychological well-being, just like the perception of social support helps to decrease the effects of stress in terms of physical and psychological problems. [16]

In relation to the physical competence, patients with satisfactory physical ability show higher levels of quality of life in the physical, psychological and in the general quality of life domains, when compared to patients with limited physical capacity. Nonetheless, there are some explanations for this. Bearing in mind another research, this is linked to various symptoms: the feeling of breathing difficulties, fatigue, body pain, nausea and fainting. [5] Some other authors support that the motor functioning is weaker in children with congenital heart diseases. [1]

Regarding the school performance item, those patients with satisfactory results revealed better quality of life levels in the psychological, environment and general quality of life domains, in contrast to patients with an unsatisfactory school performance. For some authors, an unsatisfactory schooling process can influence negatively the quality of life. [10] Hospital admissions, as well as the restriction of some activities with their peers, lead these patients to be absent from school, usually for long periods, what consequently interferes with the school success.

When differences of gender and the various domains of the quality of life were assessed only statistically significant differences in the environment domain were demonstrated. Authors sustain the idea that men can be associated to lower results in emotional and behavioural terms, while women are related to low levels of general health. [1] Furthermore, women show greater emotional support as well as a better sense of fellowship in the management of potential problems. [15]

As far as age groups are concerned, statistically significant differences were only observed in the psychological domain when comparing groups from 12 to18 and 19 to 26. Contrarily, in another study, statistically relevant differences were also detected but in what relates to motor functioning. [1]

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COMPETING INTERESTS

There are no competing interests.

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Anexo 4

Co-Autora de Um Artigo submetido para Cardiology in the Young

Living with Congenital Heart Disease (CHD): Quality of life (QOL) in early adult life

Running Title: QOL of CHD patients in early adult life

Maria Emília Guimarães Areias [1, 3], Catarina Isabel Pinto [1, 4], Patrícia Fernanda Vieira [1, 4], Marta Castro [1, 4], Isabela Freitas [1, 4], Sofia Sarmento [1, 4], Samantha Matos [1, 4], Victor Viana [2, 6], José Carlos Areias [2, 5],

1 Department of Psychology, Instituto Superior de Ciências da Saúde do Norte (CESPU), Gandra, Portugal
2 Department of Paediatrics (Cardiology), Hospital São João, Porto Medical School, University of Porto, Portugal
3 CINEICC, Centro de Investigação do Núcleo de Estudos e Intervenção Cognitivo-Comportamental, Coimbra, Portugal
4 UNIPSA, Unidade de Investigação de Psicologia e Saúde, Gandra, Portugal
5 Unidade de Investigação Cardiovascular, Porto, Portugal
6 Faculty of Nutrittion, University of Porto, Portugal

Corresponding author:
Maria Emília Guimarães Areias
Department of Psychology
Instituto Superior de Ciências da Saúde – Norte
Rua Central de Gandra, 1317
4585-116 GANDRA PRD
Portugal
e-mail: metega@sapo.pt
telephone: + 91 930 97 66
telephone: + 22 415 71 86
fax: + 22 415 71 02

Key words: psychosocial adjustment; psychiatric morbidity; congenital heart diseases; quality of life; social support;

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Abstract:

Aims: to assess the quality of life (QOL), psychiatric morbidity and the psychosocial adjustment (PSA) of adolescents and young adults with Congenital Heart Disease (CHD) and determine which variables play a role in buffering stress and promoting resilience and which ones have a detrimental effect; to investigate the situation on school performance and failures, social and family support, physical limitations and body image of these patients. **Methods:** The study enrolled 137 CHD patients (79 males), 12 to 26 years (M: 17.60 ± 3.450 years). The participants

were interviewed regarding social support, family educational style, self-image, demographic information and physical limitations. They responded to questions in a standardized psychiatric interview (SADS-L) and completed self-reports questionnaires for assessment of QOL (WHOQOL-BREF) and PSA (YSR/ASR). **Results:** We found a 19.7% lifetime prevalence of psychopathology in our participants (27.6% in females and 13.9% in males). 48% had retentions in school (M=1.61 year $\pm$ 0.82). The perception of QOL of CHD patients is better compared to the Portuguese Population in the Social Relationships and Environmental Dimensions. However, is worse in complex forms of CHD than in moderate to mild ones, in cyanotic versus acyanotic patients, in moderate-to-severe versus mild residual lesions, in patients submitted versus those not submitted to surgery, in patients with versus without physical limitations, and patients who have versus those who haven't need for medication. Social Support is very important in improving QOL of patients in all dimensions as well as academic performance. **Conclusions:** Female patients and patients with poor academic performance and poor social support refer worse psychosocial adjustment and quality of life.

Introduction:

Congenital heart disease (CHD) is defined as a malformation of the heart or the large blood vessels that develops during the fetal period. Clinically, it is classified as cyanotic or acyanotic based on the gradient of oxygen saturation in the blood [1,2].

Recent progress in early diagnosis and treatment has increased the life expectancy of patients with CHD. Today, 90% of newborns diagnosed with CHD live to adulthood, and this population is increasing at the rate of approximately 5% per year [3-6]. Advances in pediatric cardiac care have

resulted in an increasing number of adults with CHD being followed up in tertiary care centers, fact that is generating interest in adult CHD on the standpoint of a new subspecialty of cardiology. The prevalence of CHD is changing all over the world and nowadays there are more adults affected with CHD than children[7].

As survival rates improve, psychosocial issues have emerged as a critical research area.

A prominent clinical concern is patient perception of quality of life (QOL)[8], psychosocial adjustment and psychiatric morbidity.

Evaluation of health-related quality of life is becoming increasingly important for patients with CHD in view of the increase of patients who survive. Quality of life is defined as a multi-dimensional construct integrating physical, emotional, and social well-being and functioning as perceived by the individual. [9,10]

To date, studies of QOL in CHD patients have reported contradictory findings. Some studies reported that poorer QOL is related to cardiac instability [11], disease severity [12,13], motor functioning and autonomy[14], although no differences were found for the variables of gender, age or marital status [11]. Some studies found poorer psychological well-being and QOL in CHD patients compared to healthy controls [15,16], while others claimed there was no difference between the two groups. Some researchers have reported that the congenital nature of the disease leads CHD patients to have better QOL than healthy people [17,18].

Studies indicate that patients with CHD have persistent cardiac defects, a poor quality of life and psychosocial adjustment problems [19]. Moreover other studies indicate that individuals with CHD have a good psychosocial adaptation [20].

In the case of psychosocial adjustment, parents and partners tend to report more behavior and emotional problems than the patient himself.[20-22] Patients with CHD are considered to be at increased risk of psychological and emotional difficulties.[17,23] Thus, is very important to understand which variables have a detrimental effect in psychosocial adjustment and well-being of patients and which ones increase resilience and ability to adapt. The importance of our

research was that it systematically addressed the question of which demographic or clinical variables have an impact over quality of life, psychosocial adjustment, or psychiatric morbidity.

Materials and methods

Participants

The study enrolled 137 CHD patients (79 males and 58 females) with a mean age: 17.60 ± 3.450 years (range:12-26 years old) who are followed in consultation in the Pediatric Cardiology or Adult Cardiology Departments of a tertiary hospital in Portugal. We excluded patients that had not achieved a basic educational level that enables to understand and complete the written questionnaires; we only included participants who had complete medical records.

At the time of the study, two participants were married, one was divorced, and two were living in a marital union. All the other participants were single (132). Of the all participants, 20 were employed full- or part-time, 7 were unemployed, and all the others were students.

With regard to educational level, one completed the 4 first years in school, twenty the 6 first years, fifty-five the 9 first years, fifty-five completed the whole secondary education and six an university degree.

Patients' clinical files were provided by the department of Cardiology or Pediatric Cardiology. For 71 individuals, the congenital cardiac malformation was cyanotic, and for 66 patients, it was acyanotic. According to clinical files, at the time of diagnosis, 38 participants exhibited a severe form of CHD, 25 had a moderate form, and 74 had a mild form. As far as the residual lesions are concerned, 4 participants had severe residual lesions, 27 had moderate residual lesions, and 105 had mild residual lesions. The study also included the participation of 128 relatives.

The diagnosis was determined during the neonatal period for 73 of the participants, before the first birthday for 31, between the ages of 1 and 3 years for 5, between the ages of 3 and 6 years for 8 of the participants, between the ages of 6 and 12 years for 11 of the participants, and between the ages of 12 and 18 years for 9 of the participants. In several participants, the main CHD was combined with other heart diseases. Individuals with associated extra cardiac

malformations or chromosomopathies were excluded from the study. Participants exhibited the following distribution of pathologies: Transposition of the Great Arteries (20 individuals; 4 also had Ventricular Septal Defect, 1 had Aortic Stenosis, 1 had Pulmonary Stenosis, and 2 had Coarctation

of the Aorta), Tetralogy of Fallot (30), Coarctation of the Aorta (13, besides those 2 referred above), Ventricular Septal Defect (19, 1 also had Interrupted Aortic Arch, and another also had Mitral Insufficiency), Atrial Septal Defect (16 individuals; 1 had also Mitral Atresia and Pulmonary Hypertension), Atrioventricular Septal Defect (5), Aortic Stenosis (8), Pulmonary Stenosis (13), Single Ventricle (2 individuals; 1 of these individuals also had Pulmonary Atresia, and 1 had Pulmonary Stenosis), Patent Ductus Arteriosus (2), Double Outlet Right Ventricle (1), Ebstein Anomaly (3), and Pulmonary Atresia (5). For participants who underwent surgery (103), the first surgery was performed during the neonatal period for 6, before the first birthday for 35, between the ages of 1 and 3 years for 19, between the ages of 3 and 6 years for 21, between the ages of 6 and 12 years for 11 of the participants, between the ages of 12 and 18 years for 10 of the participants, and after of 18 years for 1 of the participants.

One or more of the following psychiatric disorders had been diagnosed for 27 of the participants (19, 7%) before the interview: minor or major depressive syndrome (n=14), panic disorder (n=3), anxiety disorder (n=6), or manic syndrome (n=3), and cyclothymic personality (n=1).

Assessment Instruments

The subjects were interviewed on only one occasion. A complete clinical history and demographic information were collected in a questionnaire (e.g., diagnosis, severity and category of CHD, course of illness, surgeries, presence of residual lesions, and treatment with medication) and demographic information (e.g., marital status, educational level, and occupation).

The participants also responded to a semi structured interview covering topics such as social support, family educational style, environment, self-image, functional limitations, educational background, and emotional adjustment.

A standardized psychiatric interview (SADS-L) was administered to obtain a clinical diagnosis of any psychopathologic disorders that may have existed prior to the interview. The participants completed self-report questionnaires like WHOQOL-BREF for assessment of their QOL, and YSR or ASR for assessment of psychosocial adjustment (PSA). One of their caregivers completed an observational version of the same questionnaires (CBCL or ABCL, according to the age of patients).

The WHOQOL-BREF is a self-report questionnaire that assesses subjective QOL in both healthy individuals and those with wide range of psychological and physical disorders. It is a 26-item Likert-type scale with ratings from 1 to 5. For almost all the scale items, higher scores reflect a higher QOL. However, for three items (questions 3, 4, and 26), higher scores reflect a lower QOL. The first two questions of the instrument assess general QOL. The WHOQOL-BREF also assesses four dimensions of QOL: physical (questions 3, 4, 10, 15, 16, 17, and 18), psychological (questions 5, 6, 7, 11, 19, and 26), social (questions 20, 21, and 22), and environmental (questions 8, 9, 12, 13, 14, 23, 24, and 25).

YSR or ASR are self-report questionnaires that assess behavior problems of youth or adults in the last 6 months. It is 112-item Likert-type scale for youth (YSR) and 123–item for adults (ASR) with ratings from 0 to 2. Items on the scale of youth are grouped into eight syndromes: Withdrawn, Somatic Complaints, Anxious/Depressed, Social Problems, Thought Problems, Attention Problems, Delinquent Behavior, and Aggressive Behavior. Items on the scale of adults are grouped into eight syndromes: Anxious/Depressed, Withdrawn, Somatic Complaints Thought Problems, Attention Problems, Aggressive Behavior, Rule-Breaking Behavior and Intrusive.

Procedure

Prospective participants were contacted before or after scheduled hospital appointments. The subjects were asked to participate after being fully informed of the objectives and procedures of the investigation. Those who agreed completed an informed consent form approved by the

hospital's ethical committee, which followed international conventions guaranteeing the rights of the patients.

Design

All the assessment measures were obtained on a single occasion. Clinical data were collected retrospectively using each patient's clinical record, with assistance from hospital medical staff.

Methods of Statistical Analysis

Statistical analyses of the data were performed using the IBM Social Package for the Social Sciences (SPSS), version 20.0 (SPSS, Chicago, IL, USA). The distribution of all the variables was tested. Differences for parametric variables were established using Student's t-tests, and differences for nonparametric variables were established using Mann–Whitney U tests and Chi-square tests of association.

Results

We found a 19.7% lifetime prevalence of psychopathology (27.6% in females and 13.9% in males).

48% of our participants had retentions in school (M=1.61 year $\pm$ 0.82).

There were no significant differences in QOL for presence/ absence of psychiatric diagnosis. However, QOL (Physical dimension: t= -2.926; p= 0.004) is worse in complex than in moderate-to-mild forms of the CHD as well as PSA, with patients exhibiting more internalization problems (u= 1310.000; p= 0.019) and more delinquent behavior (u= 1435.000, p= 0.042). Cyanotic patients, compared to acyanotic, have worse QOL on Physical (t= -2.575; p= 0.011) and Environmental (t= -3.149; p= 0.002) dimensions.

Patients with moderate-to-severe residual lesions had worse perception on QOL than those with mild lesions, in the Physical dimension (t= -2.379; p= 0.019). These patients also show worse

psychosocial adjustment, with more somatic complaints (u=525.500; p=0.039) and internalization problems (u=1217.000; p=0.035).

Female patients refer more somatic complaints (u= 590.500; p= 0.007), more feelings of anxiety/ depression (u= 1566.000; p= 0.002), thought problems (u= 1578.500; p= 0.001), aggressive behaviors (u= 1552.500; p= 0.001), internalization (u= 1296.000; p= 0.000), and externalization (u= 1724.500; p= 0.049) problems in PSA scales. They also show worse QOL on Environmental dimension (t= 2.856; p= 0.05).

The perception of QOL of CHD patients is better than in the Portuguese population as a whole in the Social Relationships and Environmental Dimensions, but not in the Physical Dimension. (Table 1)

Table 1. Comparison between reference values and the values presented by the participants in quality of life.

Dimensions	**Reference Values ***	**Participants of our study**	**t**	**p**
Physical	M = 77.49 DP = 12.27	M = 66.69 DP = 13.72	-15.053	**0.000**
Psychological	M = 72.38 DP = 13.50	M = 70.72 DP = 12.06	-2.562	0.100
Social relationships	M = 70.42 DP =14.54	M = 75.20 SD = 15.33	3.540	**0.001**
Environmental	M = 64.89 DP = 12.24	M = 73.16 SD = 13.14	5.768	**0.000**
General QOL	M = 71.51 DP = 13.30	M = 73.83 DP = 14.14	1.234	0.107

* For the Portuguese population as a whole

Patients submitted to surgery (N=103) have worse perception of QOL, on the Physical (t= -3.202; p= 0.002), Psychological (t= -2.949; p= 0.004) Social Relationships (t= -1.982; p= 0.049) and General Dimensions (u= 1269.000; p= 0.011) than those who were not operated (N=34). Those submitted to more than two surgeries have also worse QOL, on Physical (t= -3.541; p= 0.024) Psychological (t= -2.145; p= 0.014) and general dimensions (u= 1659.500; p=0.004). In the assessment of Psychosocial Adjustment, they also show higher scores in withdrawn behaviors (u=1335.000; p=0.036), attention problems (u= 1262.000; p= 0.014) and externalization problems (u=1209.500; p= 0.032).

Patients with physical limitations (N= 44) showed a worse perception in Physical (t= -3.123; p= 0.002), Psychological (t= -2.902; p= 0.004) and General QOL (u= 1532.000; p= 0.012) than those without PL (N= 93). They also have more withdrawn behavior (u=1454.000; p=0.006), anxiety/ depression (u=1499.500; p=0.011), delinquent behavior (u=1586.500; p=0.032) and internalization problems (u=1435.000; p=0.016).

Patients with need for medication show worse QOL only in Physical dimension (t= -2.252; p= 0.026) than those who are not medicated.

Participants with better academic performance showed better QOL on Psychological (t= 2.454; p= 0.015), Environmental (t= 2.577; p= 0.011) and General dimensions (u= 1351.000; p= 0.015). Those with poor academic performance show worse psychosocial adjustment, with more feelings of anxiety and depression (u=1312.500; p=0.013), more attention problems (u=1171.500; p=0.001) and more externalization problems (u=1190.500; p= 0.005).

Social Support is very important in improving QOL of patients in all dimensions. (Table 2) Participants with poorer Social Support show also more withdrawn behavior (u= 781.000; p= 0.000) and more social problems (u= 1011.000; p= 0.010) in PSA scales.

Table 2. Comparison of the perception of social support (more or less social support) in the various dimensions of quality of life.

	More SS (N = 109)		**Less SS (N = 28)**			
Dimensions	**M**	**SD**	**M**	**SD**	**t**	**p**
Physical	26.08	3.818	24.07	3.558	2.520	**0.013**
Psychological	23.28	2.815	21.79	2.948	2.474	**0.015**
Social relationships	12.28	1.689	11.00	2.073	3.420	**0.001**
Environmental	31.89	4.038	29.54	4.293	2.703	**0.008**
	M	**SD**	**M**	**SD**	**u**	**p**
General QOL	8.07	1.100	7.12	1.236	949.000	**0.001**

Discussion

Individuals with CHD has been increasing in adult population all over the world due to advances in early diagnosis and medical and surgical treatment [7]. Thus, the contribution of our research is important because we tested the effects of different demographic, clinical, and psychosocial variables on the perception of QOL, on psychosocial adjustment and psychiatric morbidity of CHD patients. To our knowledge, no other published study before studied so many variables as we had.

To determine the extent to which these factors enhanced resilience or had a detrimental effect on individuals with CHD, we analyzed factors such as severity of illness, number of surgeries, presence of residual lesions, presence of cyanosis, occurrence of psychopathologic disorders, education and academic achievement, size and functioning of the social support network, and physical abilities and limitations.

An intriguing finding of our study, however confirming data from other authors, is that CHD patients in the whole perceive in a better way their quality of life than the healthy population. [17, 18] That fact may be explained by the presence of some buffer variables, like family environment and cohesion, and social support.

However, when we look the different subgroups, we find that patients submitted to surgery show a worse perception on their quality of life than the whole group. These facts, more expected, may be explained by the daily life restrictions and residual side effects that limit physical performance and activity, and by the feeling of life threat and fragility, occurring in surgeries.

The comparison between cyanotic and acyanotic patients and those with moderate-to-severe or those with mild residual injuries show also a worse perception on quality of life. Complex CHD show a worse psychosocial adjustment and quality of life than moderate-to-mild forms of disease, as well.

In the literature the predictors of behavioral and emotional problems pointed are being female, having low exercise capacity, having restrictions imposed by physicians, the type of heart lesion, underwent surgery in early infancy, and a greater number of heart operations.[25-26] In this study for Psychosocial Adjustment, we found that being female, have poor academic performance, have a complex form of CHD, have moderate-to-severe residual lesions, are submitted to surgery and have physical limitations are a predictors.

The results of this study suggest that was 19.7% lifetime prevalence of psychopathology. In the others studies, the younger patients showed more psychopathology than the older patients.[23,24] On the other hand, studies on the level of psychopathology in congenital heart disease adults show conflicting results, varying from elevated levels of psychopathology to levels similar to those of peers.[28, 29]

In Portugal, there are no final data on psychiatric morbidity nationwide, although some estimation on the prevalence of psychiatric disorders in the general population could be made on the basis of partial studies.[30] Referring to those findings, we may say that CHD patients seem to show a slightly increased proneness to psychopathology.

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